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ABSTRACT

Third in a series on the prevention and treatment of developmental handicaps, this publication focuses on linkages among the network of university affiliated facilities (UAFs) and Title V Maternal and Child Health Programs. Four papers in Section I address the following developmental issues as of 1985: "Serious Viral Infection in Developmentally Disabled Persons: Threat to Normalization?" (F. Palmer, et al.); "Promises and Concerns of Technological Intervention for Children with Disabilities" (G. Vanderheiden, T. Dolan); "Community Based Services for Disabled/Chronically Ill Children and Their Families" (M. McPherson); and "The Fragile-X Syndrome" (D. Meryash). In Section II, E. Eklund updates linkages among UAFs and local Title V programs and, in a second paper, reports on a survey of inservice/preservice training needs of state Maternal and Child Health and Crippled Children's programs. The volume concludes with a listing of all papers published in the series, as well as related resource information. (JW)

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DEVELOPMENTAL HANDICAPS: PREVENTION AND TREATMENT III

A Cooperative Project
Between University Affiliated Facilities
and State MCH/CC Programs

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AAUAP
American Association of University Affiliated Programs
for Persons with Developmental Disabilities
8605 Cameron Street, Suite 406
Silver Spring, MD 20910

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The UAF Network

University Affiliated Facilities (UAFs) are programs established to train personnel in the interdisciplinary approach to the delivery of services, to conduct research and to provide exemplary services related to persons with developmental disabilities and other multiply handicapping conditions. There are 54 programs in 37 states and the District of Columbia and many of the UAFs relate to programs in other states.

The American Association of University Affiliated Programs for Persons with Developmental Disabilities (AAUAP) is the national association of UAFs.

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Project Steering Group:

Allen C. Crocker, Chairman
Director, Developmental Evaluation Clinic, The Children's Hospital, Boston, Massachusetts
Alfred Healy, Vice-Chairman
Director, Division of Developmental Disabilities, University Hospital School, The University of Iowa, Iowa City, Iowa
Herbert J. Cohen
Director, Rose F. Kennedy Center, Albert Einstein College of Medicine, Yeshiva University, Bronx, New York
Antoinette Eaton
Associate Medical Director, Ambulatory Services, Columbus Children's Hospital, Columbus, Ohio
Judson Force
Chief, Division of Crippled Children's Services, Mental Retardation and Developmental Disabilities Administration, State Department of Mental Health & Hygiene, Baltimore, Maryland
William Hollinshead
Chief, Division of Family Health, Department of Health, Providence, Rhode Island
Phyllis R. Magrab
Director, Child Development Center, Georgetown University, Washington, D.C.

Sponsors:

Marvin Fifield
Director, Developmental Center for Handicapped Persons, Utah State University, Logan, Utah
John C. MacQueen
Director, Iowa Specialized Child Health Services, The University of Iowa Hospitals & Clinics, Iowa City, Iowa

Project Coordinator:

Elaine M. Eklund, Associate Director, American Association of University Affiliated Programs, 8605 Cameron Street, Suite 406, Silver Spring, MD 20910

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Introduction

In this, the third edition of the Developmental Handicaps series, the focus is on linkages that exist among and between Title V programs and, specifically, between the network of University Affiliated Facilities (UAFs) and state Maternal and Child Health and Crippled Children's programs. The impetus for cooperation derives not strictly from the intent of Title V legislation, but from the mutual desire of the programs involved to improve services to disabled and chronically ill children and their families.

Toward this end, this publication contains four state of the art papers on current topics of concern in the Title V arena and, in the second section, explores the current status of linkages between UAFs and Title V programs in the states. The latter is followed by a review and discussion of the results of a 1983 survey of training needs of Title V personnel from which current working relationships can be traced.

The issue papers start with a critically timely topic: "Serious Viral Infection in Developmentally Disabled Persons: Threat to Normalization?" The authors review current knowledge of certain infections as it relates to developmentally disabled persons and focus on risk of transmission and infection control.

The paper responds to the controversy which has grown as the prevalence of private and mandated public developmental programs has increased in the past decade and which has been heightened by the media attention paid to certain infections in the nondisabled population such as genital herpes infections, hepatitis, and acquired immune deficiency syndrome (AIDS).

The second paper looks at the "Promises and Concerns of Technological Intervention for Children with Disabilities," particularly the application of computer technology to both therapy and training in independent living skills

for children and adults with chronic handicapping conditions. The paper reviews the potential benefits of the developments and explores associated concerns.

"Community Based Services for Disabled/Chronically Ill Children and Their Families," begins with a philosophical/historical review of the concept of community based services and describes several recent or current collaborative and networking projects contributing to the further development of community based services. The last paper, on the "Fragile-X Syndrome," concerns an inherited genetic condition which is a relatively frequent cause of mental retardation (estimated to affect one out of every 2,000 males among all major races and cultures in the United States) which has only been recognized as a distinct entity in the past decade.

In the second section, readers will find an update on the state of working relationships. Some of the activity is "new" where projects have been initiated in the past year. A number of UAF-state liaisons are reported. Some of those represent first-time relationships between the UAF and the state Title V program. Others have longer histories, but the vitality of the relationships has been maintained as evolving needs in training or service are dealt with. New or old, the descriptions of cooperation are offered as examples of ways in which programs can collaborate for their mutual benefit and in support of disabled and chronically ill children and their families.

Silver Spring, Maryland — Elaine M. Eklund

November, 1985

I.
DEVELOPMENTAL ISSUES:
THE STATE OF THE ART 1985

SERIOUS VIRAL INFECTION IN DEVELOPMENTALLY DISABLED PERSONS: THREAT TO NORMALIZATION?

Frederick B. Palmer, M.D.
The Kennedy Institute for Handicapped Children (UAF)
Baltimore, Maryland

Philip Ziring, M.D.
Morristown Memorial Hospital
Morristown, New Jersey

Bruce K. Shapiro, M.D.
The Kennedy Institute for Handicapped Children (UAF)
Baltimore, Maryland

Infection in the developmentally disabled infant and child has become an extremely controversial topic as the prevalance of private and mandated public developmental programs has increased in the last ten years. Concern about contagion in community developmental centers has led to an avalanche of publicity, waves of anxiety and a variety of legal challenges. The excitement is heightened by the media attention paid to certain infections in the nondisabled population such as genital herpes infections, hepatitis, and acquired immune deficiency syndrome (AIDS). This paper is intended to briefly review current knowledge of certain infections as it relates to the developmentally disabled. Emphasis is placed on risk of transmission and infection control. It is hoped that this information will provide those caring for developmentally disabled persons with an improved understanding of contagion and contribute to effective and dispassionate efforts of infection control.

Cytomegalovirus Infections

Cytomegalovirus (CMV) is the most common agent known to be responsible for congenital infection. It is a DNA virus like the herpes viruses. Growth of CMV in human fibroblasts results in a characteristic cytopathic effect of cell

enlargement and intranuclear inclusions. These inclusion cells often are visible in the urinary sediment of infected individuals providing a simple (but only partially sensitive) method for clinical diagnosis. Until the virus had been grown in tissue culture in the 1950's, CMV infection was rarely diagnosed. With the widespread use of these techniques and serologic methods, CMV infection is now commonly and readily diagnosed and the virus is recognized as ubiquitous in the community (Krugman & Katz, 1981).

Clinical Syndrome

Intrauterine CMV infection is the most common congenital infection, occurring in up to 2.3% of live births. Ninety-five percent of congenitally infected infants are asymptomatic and have no sequelae. Those with symptoms experience a wide range of problems including neonatal hepatitis, purpura, and microcephaly. The most common clinical feature may be intrauterine growth retardation including microcephaly. Other findings include intracranial calcifications, deafness, chorioretinitis, and various congenital anomalies. The central nervous system damage and resultant developmental disability are of greatest concern. Neurologic sequelae range from profound mental retardation, cerebral palsy and seizures, to communication disorder, hyperactivity, learning disability, and sensorineural hearing loss (Hanshaw, 1970). Infants with congenital CMV infection may continue to excrete the virus in urine and body secretions such as saliva for a prolonged period of time, months to years.

There is a vast range of clinical presentation of postnatal CMV infection ranging from the asymptomatic to severe disseminated manifestations. Postnatal primary infections in otherwise healthy infants, children, and adults are most often innocuous and commonly subclinical. Mild respiratory symptoms possibly with a rash are not uncommon. More severe infections including hepatitis and an infectious mononucleosis-like picture with fever, rash and polyarthritis occur. The severe forms of infection are more likely in the debilitated or

immunocompromised individual. Transmission of CMV through blood transfusion or organ transplant is a continuing difficulty in the debilitated patient. CMV often persists in the body and is excreted in the urine and other fluids years after the mildest postnatal infection. A secondary or recurrent CMV infection can result from reexposure to an antigenically different strain or reactivation of latent virus at a time of physiologic stress such as surgery, pregnancy, illness, or the use of immunosuppressive drugs. It is now apparent that symptomatic intrauterine CMV infection occurs most frequently with primary maternal infection and not as a result of secondary infection (reexposure or reactivation) (Stagno, Pass, Dworsky, Henderson, Moore, Walton & Alford, 1982).

Transmission

Considerable concern has been expressed by parents, teachers, physicians, and public health officials about the risk to pregnant or potentially pregnant women exposed to CMV. The focus of this concern is usually the presence of a known virus excretor in a day care or developmental center. In order to understand the issues which determine risk and to develop recommendations to control risk, it is helpful to briefly review the epidemiology of CMV infection.

CMV virus is ubiquitous in the community. It is not highly contagious and requires intimate or close interpersonal contact with virus excretors to be transmitted. Thirty five to 90% of females of childbearing age are seropositive for CMV, that is, they have evidence of previous primary infection. At present, seropositivity is much more common in low socioeconomic classes, in blacks, and in situations of overcrowding which increase the likelihood of transmission (Krugman & Katz, 1981; Hutto, Ricks & Pass, 1985).

The increasing use of child day care and developmental centers appears to be altering the epidemiology, probably by providing frequent opportunity for

close intimate contact between infants, some of whom are virus excretors. In such settings, transmission through urine and saliva is probably of greatest importance. An evaluation in a day care center in Alabama composed primarily of white middle income families showed that 57% of enrolled children aged birth to 5 years were excreting CMV in their urine or saliva. Further, in the age group 12-24 months, almost 80% were excretors. This contrasts with a comparison group of preschool aged children not attending a day care center where the virus excretion prevalence was only 8% (Pass, Hutto, Reynolds & Polhill, 1984). Although absolute prevalence of excretion will vary from center to center or class to class, it is apparent that the increasing use of out-of-home care for normal and handicapped children may increase the risk of infection in pregnant women by increasing the number of infants and preschoolers who are excreting the virus. This increased risk appears to be independent of socioeconomic class or race.

The degree of risk this presents to the pregnant caretaker is unclear. Correlations have been demonstrated between CMV infections in infants and children and subsequent seroconversion of mothers and other adults in the household. However, the magnitude of increased risk for CMV infection in a parent or day care worker or, more importantly the increased risk for intrauterine infection, is uncertain.

These uncertainties have a bearing on policy regarding day care or developmental center attendance of normal or handicapped infants known to be excreting CMV. Since CMV is ubiquitous in day care centers, almost any center is likely to have children enrolled who are excretors. It is illogical to exclude known excretors when there may be many other unknown excretors in the same center. Since the prevalence of CMV excretion in developmental centers is likely to be no higher than that in day care centers for normal children (Jones, Duke-Duncan & Yeager, 1985), the same argument should apply to the exclusion of

infants with known congenital infection. This is especially important as exclusion could deprive the child of needed developmental programming. The unavoidable risk of repeated exposure to children in the general population who are unrecognized virus excretors probably far outweighs the risk from a child with known congenital CMV (Hutto, et al., 1985).

It remains likely that the most effective method of reducing the risk of transmission of CMV is careful handwashing after contact with all infants, especially contact with saliva, urine, or other bodily secretions (Hutto, et al., 1985; Pass & Kinney, 1985). The importance of such simple sanitation measures needs to be emphasized to all child care workers and center administrators. Currently, routine serologic screening of child care workers for CMV seronegativity (and thus risk for primary infection) cannot be advocated because of limited knowledge about actual risk and tremendous expense. Whether transfer of the worker to another setting would significantly influence risk is not known (Hutto, et al., 1985; Pass & Kinney, 1985).

Summary

CMV is the most common known agent in congenital infection. The virus produces a wide range of symptomatology in congenitally infected infants ranging from inapparent infection to severe disability, fetal loss, or infant death. Postnatal infection is extremely common and usually mild or inapparent in otherwise healthy individuals. The increased use of day care and developmental centers may be altering the epidemiology of infection in young children and may result in increased opportunity for exposure to women of childbearing age. Simple infection control measures are felt to be the best method for prevention of transmission.

Accurate data quantitating risk to mothers and child careworkers is necessary. With such information, rational policy to reduce transmission can be

developed. Based on current information, it is illogical to exclude infants from day care or developmental centers based on CMV excretion (Hutto, et al., 1985; Pass & Kinney, 1985; Jones, et al., 1985).

Herpes Simplex Virus Infections

Surveying the media reveals the strong emotional feelings associated with Herpes Simplex Virus (HSV) infection. Headlines such as "Herpes Epidemic Rages," "Mother's Kiss Causes Infant's Handicap," and "Parents Vow Boycott if Herpes Child Attends Class" indicate the fear, concerns, and misinformation which the general population possesses. This section reviews existing data about HSV to provide information for those who are likely to deal with children who have been handicapped as a result of HSV infection.

The Virus

HSV is a large complex virus which is related to cytomegalovirus and varicella zoster virus. All members of this class have double stranded DNA in their core and consequently have the potential for being incorporated into cellular DNA. In addition to causing cell death by overwhelming biosynthesis of infectious progeny, the HSV genome can be totally or partially incorporated into cellular DNA, with continuing cell survival. This latent (nonproductive) infection can be reactivated at a later time by a variety of stimuli such as fever, sunshine, trauma, menses or psychic stress. Reactivation manifested by local vesicular eruptions, e.g., the common "cold sore," is usually not associated with constitutional symptoms. The mechanisms of reactivation are unknown. It is also not known whether the virus is ever cleared from the host organism.

HSV is divided into two subtypes based upon epidemiology and biologic effects (Nahmias & Roizman, 1973). Primary HSV1 infection occurs in young

children. Ninety to 95% of HSV1 infections are not associated with clinical disease. Symptomatic HSV1 may present with lesions of mucous membranes (lips, oropharynx, eye, genitals), lesions related to direct innoculation (whitlow), or meningoencephalitis.

HSV2 is found primarily in adolescents, young adults, and neonates. It too is commonly associated with subclinical disease. HSV2 causes genital infections but non-genital infection can be seen. Neonatal meningoencephalitis is primarily caused by HSV2 and has been associated with maternal infection. Approximately 1% of pregnancies have cytologically detectable primary or recurrent herpetic genital lesions.

Transmission

The usual route of transmission is via direct, intimate contact. HSV1 is transmitted in an unknown fashion, although respiratory spread is postulated. HSV2 is acquired through venereal contact. However, HSV1 is found in approximately 10% of genital infections and HSV2 has been found in the pharynx. Genital herpes in a prepubertal child should raise concern of sexual abuse.

Although asymptomatic infection is the usual result of HSV infection, those with deficient immunologic defense such as immunosuppressed patients or neonates, represent special risks. The degree of risk is unknown and this is reflected in the American Academy of Pediatrics (AAP) recommendations.

In the case of "cold sores," not kissing newborn infants or infants with eczema is advised. The decision to relieve a person with a "cold sore" from direct patient care because of potential risk of infection must be balanced against the risks of compromising care by excluding personnel who are essential for the operation of the unit. Further, there is a 9.6% rate of recovery of HSV from saliva samples of asymptomatic personnel. Persons with herpetic whitlow (HSV infection of the finger) should not work in direct patient care activities.

Personnel with genital HSV infection require no special procedures (AAP, 1982).

To date the AAP has no official policy regarding techniques for dealing with recurrent HSV infection in a congenitally infected child. Intermittent shedding of virus may occur years after infection. The degree of infectivity of patients with recurrent lesions is unknown. In the immunologically normal population, HSV is a mild disease which is usually asymptomatic. Whether additional isolation is needed for nonhospitalized children will require better understanding of methods of transmission. The likelihood of significant disease resulting from healthy children (or care takers) being together in a classroom with a child who suffered neonatal infection is remote. At present it seems prudent to cover active skin lesions with a sterile dressing to limit lesion-to-skin contact and decrease the likelihood of bacterial superinfection.

Symptomatic HSV infections are estimated to occur in 1/7,500 births. This incidence figure is subject to question but it is clear that the fetus' unique relationship with the mother and its immature immunological system permits other modes of HSV transmission to occur. Maternal viremia may allow virus to pass through the placenta to the fetus, although transplacental infection is uncommon. Intrapartum acquisition is felt to be the major route of transmission of neonatal HSV infection. It can occur during passage through an infected birth canal or via ascending infection through ruptured amniotic membranes. Approximately one half of children born to women with genital herpes at or near term develop an infection and approximately half of those have severe or fatal disease (Kibrick, 1980).

Clinical Syndromes

Several preliminary reports have suggested that antenatal HSV infection is associated with increased rates of abortion, preterm birth, and congenital malformations. Mothers who contracted HSV prior to 20 weeks gestation had a threefold increase in spontaneous abortion (Nahmias, Josey, Naib, Freeman,

Fernandez & Wheeler, 1971). Preterm birth approximated 50% in infants of mothers who evidenced HSV infection during pregnancy (Whitley, Nahmias, Visintine, Fleming & Alford, 1980). Several reports of microcephaly and microphthalmia associated with transplacental infection have been described (Hanshaw & Dudgeon, 1978).

Neonatal HSV disease is primarily the result of HSV2 infection. Most of the mothers are asymptomatic and without obvious signs of HSV at the time of delivery (Whitley, et al., 1980). Although primary HSV in the mother is postulated to be more likely associated with neonatal disease (Kibrick, 1980), serologic studies have shown that prior maternal HSV exposure is common in cases of neonatal infection (Whitley, Nahmias, Soong, Galusso, Fleming & Alford, 1980a). Therefore, neonates may be infected as a result of a primary or reactivated maternal infection.

Intrapartum acquisition is usually insidious, not becoming evident until 6-12 days or as late as one month. Neonatal HSV may assume three forms--localized to eyes, skin, or mucosa; localized to central nervous system (CNS); or disseminated visceral disease affecting lung, liver, or other organs (including CNS). Maternal antibody status does not predict which form of HSV disease develops nor does it influence long term survival and morbidity of the infected infant (Whitley, et al., 1980a).

Most babies with HSV disease (80%) present with vesicular eruptions. The appearance of vesicles is ominous as the majority of infants progress to disseminated or CNS disease. Diagnosis may be delayed in infants who are at home and in whom the significance of the vesicles is not apparent. The appearance of vesicles may initially escape detection in hospitalized, ill prematures. Finally, a substantial number of infants do not present with vesicles and thereby constitute a major diagnostic challenge.

Details of other HSV infections are outlined elsewhere (Krugman & Katz, 1981).

Treatment

Mortality rates in neonatal HSV infections range from 30% for localized CNS disease to 80% in disseminated forms. Profound CNS handicaps -- microcephaly, seizures, quadraparesis, profound cognitive deficits, and post inflammatory eye disease -- are the rule with only 10-15% of neonates surviving without residua. Localized disease carries a better prognosis for life but is still associated with a rate of handicap of approximately 15%. Adenine arabinoside has become the treatment of choice for neonatal HSV infection and has decreased the mortality from 74% to 34%. The most dramatic effects have been noted in disseminated and localized CNS forms of the disease where mortality was decreased from 85 to 57% and 50 to 10%, respectively. The difference in the morbidity is less impressive for disseminated disease (14 vs. 8%) but more encouraging for localized CNS disease (50 vs. 17%) (Whitley, et al., 1980a).

Adenine arabinoside is a purine analogue and, while it does not show substantial long term neurologic toxicity in adults, its effects on the developing CNS must be viewed with caution. It is possible that the "cure" may result in neurologic dysfunction. However, the number of children treated is small and length of follow-up too short to satisfactorily resolve this issue. Given the extreme mortality and morbidity and the lack of a proven alternative (trials are underway to determine the efficacy of acyclovir, another nucleotide analogue) there is little choice at the current time.

The treatment of localized eye infection requires the supervision of an ophthalmologist and is urgent. Meningoencephalitis in patients outside the neonatal period is beyond the scope of this review. The remaining clinical syndromes are usually managed supportively (Krugman & Katz, 1981).

Summary

People who deal with handicapped children develop a biased view of HSV. Instead of seeing a disease which is usually mild or asymptomatic, they see the results of intrapartum acquisition of HSV. The profound sequelae associated with this form of HSV infection tend to overshadow the broader picture and lead to unfounded associations and emotional postures.

The most effective way of dealing with handicap resulting from HSV infection is to prevent its occurrence. The method of transmission of genital HSV to the immunologically immature neonate is well established. Expectant mothers with a history of HSV infection should be examined clinically and cytologically on a regular basis to determine if lesions are present. If lesions present near term or if genital HSV lesions are noted in a woman whose amniotic membranes have ruptured less than 4-6 hours, then cesarian section should be considered.

Although adenonine arabinoside has decreased the mortality of neonatal HSV infection, the mortality is still unacceptably high in some groups and morbidity remains substantial.

The likelihood of significant disease resulting from children (or caretakers) being together in a classroom with a child who suffered neonatal infection is remote if the children (or caretaker) are immunologically competent and lesion-to-skin contact is avoided.

Hepatitis

Hepatitis is a viral illness affecting the liver which is prevalent throughout the world and has been responsible for serious morbidity among all age groups and socioeconomic strata (Krugman & Katz, 1981). It is a disease

which has had a very special significance for mentally retarded persons in the United States during the past fifty years, first as an endemic illness in public residential facilities throughout the country, and more recently as a public health issue regarding the rights of mentally retarded persons, who are carriers of Hepatitis B surface antigen, to live in the community. It will remain a problem until there is far better education of the public and mental retardation professionals regarding the natural history and means of prevention of this disease. There is no easy solution for this problem in sight, and it is a major responsibility for professional staff from all disciplines to be fully informed about the nature of this disorder, its management, and means of prevention.

Hepatitis A and Hepatitis B

Two distinct forms of viral hepatitis were not recognized until the World War II era, when infectious Hepatitis (now referred to as Hepatitis A) was distinguished from Serum Hepatitis (now referred to as Hepatitis B) by some of its clinical features, such as its shorter incubation period and mode of spread. It was not until the last twenty years that a quickening pace of scientific investigation led to a number of major breakthroughs in our understanding of other important features of these diseases, their long term consequences, and to the development of effective means of prevention.

Hepatitis A, caused by an RNA virus, is now known to be an acute illness of widespread distribution throughout the country, especially prevalent in children as an inapparent or subclinical infection, and readily spread to susceptible individuals through the "fecal-oral" route. It is generally not spread by contaminated blood or needles, is characterized by a carrier state, and is usually a self-limited illness followed by complete recovery. Prophylaxis of exposed persons with ordinary pooled gamma globulin is often helpful in preventing serious clinical illness. Preexposure prophylaxis is also recommended under circumstances of travel to areas of the world where Hepatitis A is highly

endemic (MMWR, 1985c). A vaccine for prevention of Hepatitis A is currently under development, but is not yet available for general use.

Hepatitis B, caused by a more complex DNA virus (HBV), is of more limited prevalence in the United States, where it is of special concern for certain high risk groups, such as intravenous drug abusers, male homosexuals, and persons in the health professions. The disease is generally spread by contact with blood contaminated by HBV. Although less likely, HBV may also be transmitted by mucosal contact with other infective body secretions such as saliva or semen. The disease is not spread by exposure to the stool of a carrier (unless the stool is contaminated by blood) and ordinary immune globulin is only of limited efficacy as a prophylactic agent. Ninety percent of individuals contracting the disease recover fully and become immune to the disease by developing antibody to Hepatitis B surface antigen. Six to ten percent of individuals acquiring Hepatitis B do not develop antibody but remain chronic carriers of HBV and have the capacity, under circumstances of very intimate exposure, to transmit the disease to susceptible individuals. A small minority of such carriers are more highly contagious than the remainder, and are identified by demonstrating e-antigen in their blood. These long term carriers have significant subsequent morbidity with more than 25% developing chronic active hepatitis and, in some, cirrhosis. Furthermore, there is an association between the HBV carrier state and the occurrence of primary liver cancer later in life (MMWR, 1985c).

Susceptible persons who have lived in residential facilities for the mentally retarded and who may be intimately and repeatedly exposed to infected persons are commonly infected by HBV because of the limited hygienic practices in such settings. For example, while the prevalence of HBV carriage in the general community is approximately three per thousand, in institutions for mentally retarded persons it is often as high as 30%, with many of these

individuals also being positive for e-antigen.

The increasing trend toward normalization in the United States has seen more mentally retarded persons continuing to live in the community. They are joined in the community by persons who have been deinstitutionalized from residential centers and may be HBV carriers. The result often has been great anxiety on the part of parents of retarded persons that their children would become infected with Hepatitis B by these carriers. Similarly, staff in special education programs, workshops, and group homes may be fearful of contracting Hepatitis through their occupational exposures. Furthermore, the public at large is concerned about the possibility of the spread of Hepatitis throughout the community.

In actual fact, most of these fears do not seem to have been justified by experience. Available data from public schools in which mentally retarded children who are carriers of HBV are integrated with mentally retarded children susceptible to Hepatitis have failed to demonstrate significant spread (Williams, Weber, Cullen & Kane, 1983). Similarly, where adequate hygienic practices are carried out (proper disposal of materials contaminated by body fluids, adequate handwashing), there has been little documentation of spread of Hepatitis B, even under the more intimate conditions of group home living.

Prevention

The recent widespread availability of a safe and effective Hepatitis B vaccine should lead to considerable reduction in the risk to susceptible clients and staff posed by their exposure to persons who are chronic carriers of HBV. A full series of three innoculations of this vaccine has been demonstrated to induce immunity in 95% of individuals. Side effects have consisted primarily of local, transient soreness at the injection site and occasional low grade fever. The vaccine is prepared from plasma of individuals who have had Hepatitis B and are carriers of the antigen, but the procedures utilized in the preparation of

the vaccine are believed to destroy all known infectious agents, including the virus which is believed to be responsible for causing AIDS.

It has been recommended, therefore, by the Immunization Practices Advisory Committee of the Centers for Disease Control of the Public Health Service that persons who may be intimately exposed to carriers of Hepatitis B, especially carriers demonstrated to be e-antigen positive, be given this Hepatitis B vaccine (MMWR, 1985c). Such individuals currently deemed to be at higher risk than the general population would include susceptible individuals being newly admitted to residential facilities for mentally retarded persons, health workers who have frequent contact with materials from carriers, and direct care and therapy staff who have close and direct contact with carriers whose behavior makes transmission possible. This includes community group home residents and staff as well as those from larger residential facilities. In view of the present high cost of the vaccine, persons who are candidates for the vaccine may choose to be screened serologically for evidence of existing immunity to Hepatitis B, although only approximately 5% of individuals in the general population have such evidence of prior infection. Immunization of susceptible individuals as currently recommended should effectively eliminate the already low risk of exposure to mentally retarded persons who are carriers and allow for their full integration into community based programs.

Summary

Hepatitis B infection has represented a special problem in the day-to-day management of programs serving persons with mental retardation. Thorough education of the public and of mental retardation professionals regarding the natural history and means of prevention of this disease can go far in allaying the fear surrounding this problem and limit the risk of spread of the infection. Proper utilization of the Hepatitis B vaccine can effectively eliminate even the

small risk these clients pose to others in their community.

Acquired Immunodeficiency Syndrome

Acquired immunodeficiency syndrome (AIDS) has become a familiar term to many adults as a result of its continually increasing incidence, high prevalence in homosexuals, and sensational media coverage. Recently a pediatric form has been recognized (Rubenstein, Sicklick, Gupta, Bernstein, Klein, Rubenstein, Spigland, Fruchter, Litman, Lee & Hollander, 1983), raising issues of transmission to others similar to those raised by CMV, Hepatitis B, and HSV.

AIDS, as recognized in adults, is a syndrome of severe, generalized immunologic dysfunction. Patients are incapable of mounting effective immune responses to foreign antigens and yet have inappropriate formation of auto-antibodies and immune complexes, and therefore show autoimmune disease. The deficient immune responses result in severe, recurrent, and opportunistic infection. Although the natural history of AIDS is as yet incompletely understood, it appears to be irreversible with a uniformly poor prognosis. Seventy-six percent of all those diagnosed before January, 1983, had died by August, 1985 (CDC, 1985).

Recognized risk factors for AIDS include male homosexuals, drug addicts, heterosexual prostitutes, patients with hemophilia and others receiving blood or blood product transfusions, and certain young children (Shannon & Ammann, 1985). The etiologic agent appears to be human T-cell lymphotropic virus (HTLV-III). HTLV-III has been isolated from blood, semen, and saliva of infected individuals. Information suggests that transmission occurs through intimate contact or contaminated blood or blood products. Routine contact such as casual kissing, hugging, or other social contact has not been shown to result in virus spread. The experience with AIDS in children suggests that transplacental

infection also occurs.

As of September, 1985, 165 cases of AIDS in children under age 13 had been reported (CDC, 1985). Seventy percent were children with parents who either had AIDS or were at increased risk for AIDS. Twenty percent apparently contracted AIDS through blood or blood product transfusion. Incomplete information is available regarding risk factors for the parents of the remaining 10% of infected children. Infants with apparently transplacentally acquired AIDS usually present with interstitial pneumonitis, hepatosplenomegaly, developmental delay, and failure to thrive, many before 3 months of age. They soon develop severe infections. As in adults the prognosis is poor. Of the 165 reported pediatric cases, 113 have died as of September, 1985. An AIDS prodrome occurs and is characterized by the immunologic and clinical features but without recognized opportunistic infection. This prodrome, AIDS-Related Complex (ARC), will probably be recognized with increasing frequency as physicians' familiarity with the AIDS spectrum grows. Whether all with ARC go on to develop full blown AIDS with its bleak prognosis is unknown. Better knowledge of the natural history of AIDS and ARC in children should emerge in the next few years.

Recently infants and children with AIDS have been shown to have a wide variety of neurologic and developmental abnormalities (Belman, Ultmann, Spiro, Rubenstein, Horoupian, & Cohen, 1984; Ultmann, Belman, Ruff, Novick, Cone-Wesson, Cohen & Rubenstein, in press). These abnormalities may be due to many factors including recurrent infection, nutritional compromise, prolonged hospitalization, and deprivation, as well as direct involvement of the CNS. Such neurologic and developmental abnormalities will result in recommendation for placement in various developmental programs. Furthermore, the disordered social and family circumstances of many AIDS infants will require a wide range of public services such as foster care, social and protective services, and

health department involvement. Each of these needs increases the contact with adults and heightens anxiety about transmission of AIDS.

There are well-established guidelines for the care of hospitalized patients with AIDS and for precautions to be taken by patients with AIDS (Conte, Hadley & Sane, 1983). These guidelines are designed to eliminate the transmission of virus by contact with contaminated needles, blood products, or bodily secretions. When these guidelines are followed, transmission of AIDS to hospital workers caring for AIDS patients has not been a problem. In fact, prior to establishment of guidelines, there is no evidence of spread of AIDS to health care workers through caring for AIDS patients. Since casual contact is not felt to lead to transmission of AIDS, it is reasonable to conclude that routine contact in a developmental center or foster placement also should not be a problem. In addition to careful attention to handwashing, precautions to avoid blood contact should be taken. After accidents with bleeding, surfaces contaminated with blood should be cleaned with a 1:10 solution of bleach in water. Toothbrushes which can become contaminated with blood should not be shared (MMWR, 1985a). Dentists, dental hygienists, and others involved in dental care may have to take particular precautions due to their close exposure to saliva and blood.

Neurologically impaired infants and children with AIDS who cannot control saliva and other bodily secretions pose a theoretical risk for transmission. Decisions regarding their management should be made after discussion with the child's physician, parents or guardian, and school personnel.

The Centers for Disease Control have recently circulated recommendations for management of infants and children with AIDS in foster care and educational settings (MMWR, 1985c). Those caring for such children should review these recommendations carefully.

As with other infections already discussed, denial of services to the child

and family afflicted with AIDS seems an unnecessary additional burden to bear. Calm attention to accepted practices of infection control and an avoidance of sensationalism will result in safe delivery of necessary services to the child and family with AIDS.

IMPLICATIONS FOR UAFs AND TITLE V AGENCIES

The four viral infectious diseases discussed in this review present a series of common problems, particularly in relationship to the safe and optimal delivery of developmental services to children who have suffered the chronic sequelae of these diseases. Title V agencies and UAFs may be positioned to offer solutions to some of these problems.

1. What is the risk of exposure to infected infants and children? There is substantial epidemiologic information about the mode of transmission of each virus but there is little specific information about how this knowledge can be applied to issues of transmission in developmental and daycare centers. Does employment at a center servicing infants and children place a woman at significantly increased risk for developing CMV and passing it on to her fetus? In the cases of HBV and AIDS, where the risk of transmission is certainly low, it is nevertheless important to develop systems of ongoing data collection and monitoring to identify those specific situations in which there is higher risk. This kind of health data will be most valuable when collected across several sites which reflect the diversity of care provided in a region rather than in a single site. State Crippled Children's or Maternal and Child Health agencies, ideally in cooperation with educational agencies, should be able to assist in the organization, direction, and funding of such projects. The information derived will be necessary to develop more specific guidelines to reduce risk.

2. The development and implementation of model standards. Whatever the public health solutions to the prevention of infectious disease transmission, their success will depend upon the consistency with which they can be implemented. A collaborative effort between state health and education agencies will be necessary to design, implement, and monitor practical guidelines for infection control. The establishment of guidelines based on improved knowledge of actual risk will be of great benefit in reducing the anxiety and sensationalism surrounding children with these conditions.

3. Education. Current information about transmission indicates low levels of risk when proper sanitation and hygiene procedures are followed. Yet this knowledge is not widely disseminated, a fact which contributes to anxiety among parents, teachers, caretakers, and community officials. Many of the UAFs, in their mission of community outreach, are well suited to develop and implement training modules which provide up-to-date information regarding natural history, risk of contagion, and methods of infection control. The potential audience for such training is extremely broad. It should include trainees in professional programs, workers in the community (inservice programs), family members of the developmentally disabled, and the general public.

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PROMISES AND CONCERNS OF TECHNOLOGICAL INTERVENTION FOR CHILDREN WITH DISABILITIES

Gregg C. Vanderheiden, Ph.D.
Terrence R. Dolan, Ph.D.
Waisman Center UAF
University of Wisconsin
Madison, Wisconsin

Introduction

The past decade has been marked by significant technological advances in the biomedical and behavioral sciences, particularly in those areas benefitting from developments in computer technology. Although many of these advances have been in research and diagnosis, the advent of the portable microcomputer has introduced new ways to apply computer technology to both therapy and training in independent living skills for children and adults with chronic handicapping conditions. These new computer-related tools and systems have the potential to improve significantly the communication skills of handicapped persons, to enhance their independence and productivity, and to increase their ability to participate in normal societal activities (Vanderheiden, 1981).

Although new technological developments offer significant opportunities for realizing the potential of persons who have chronic handicapping conditions, they also raise a number of concerns. It is important to address these concerns, and to emphasize that if individuals with disabilities are truly to benefit, technological advances in the rehabilitation field must keep up with and be coordinated with those in the society at large.

This paper addresses the potential contributions of recent technological developments to the field of rehabilitation, discusses several concerns and reservations to be considered in applying these technologies, and offers some

thoughts about the implications of these developments for UAFs and Title V programs.

Potential of the New Technology

The potential benefits of the new technologies to the lives of disabled children fall into three categories:

1) the facilitation and automation of some therapy and educational activities;

2) the provision, restoration, or extension of an individual's physical abilities; and

3) the provision of opportunities for greater participation in the regular activities of society.

Facilitation and Automation of Therapy and Education

One use of technology in the rehabilitation process is in the area of therapy and education. Here, technology is used to facilitate the process of developing new skills and competencies (either physical or cognitive) in the disabled individual. Although there have always been aids and devices to facilitate therapy and education, newer technologies, particularly the microcomputer, are providing opportunities for newer, more effective, and less clinician-intensive therapy strategies. In addition to affording better diagnostic techniques, these technologies can also serve as "therapist-extenders." This means either that a therapist can work with many clients

> A deaf child trying to learn to speak has difficulty in imitating the sounds of his teacher. He can see the teacher's lips and feel the vibrations of her throat, but he cannot see what she does inside her mouth. "Ga" and "ka," for example, consist mostly of tongue movement at the back of the mouth. Using a computer simulation showing a cutaway view of the inside of the mouth and throat, the deaf boy can see what the entire mouth is doing, as well as see how the flow of air, movement of the tongue and phonation are related in time for different words.

concurrently (each of them with his/her individual computer or computer-driven

therapy system), or that a client can augment his/her direct contact with additional automated-therapy sessions. Use of a computer to help an individual in developing a vocabulary or in extending his/her command of syntax would be examples of applications in this area. Similarly, the use of video games which require eye-hand coordination and attentive mental activity can be used in cognitive rehabilitation programs. Development of memory, visual-spatial and attending skills, as well as the learning of specialized input routines, can also be facilitated using computers. Computer-aided diagnosis, where the stimuli are presented by the computer, and the responses of the client recorded and tracked by the computer, can lead to more consistently administered tests and to tests which are more easily taken by individuals with severe disabilities.

> A non-vocal young girl in a rural community doesn't have access to regular language therapy. Using a program on her Apple computer, she works with her older (non-handicapped) brother practicing her communication skills as part of a communication development program laid out by her visiting speech therapist, whom she sees once a month.

One advantage of computers in this role is that they are infinitely patient. Unlike human therapists, they can work one-on-one with disabled individuals for very extended periods of time without fatiguing or losing concentration. They can also help monitor client progress and continually modify what they present to the client in a consistent and controlled fashion.

One drawback to the use of computers as auxiliary "therapists" at the present time is that their "intelligence" and effectiveness in unsupervised situations is limited. The effectiveness of computers in these applications is limited by both a lack of knowledge and understanding of various handicapping conditions and the current state of artificial intelligence systems. As

knowledge in these fields grows, there will be more refined techniques and new applications. These applications might include the provision of language stimulation and modelling for young children, self-based exercise for skill development in brain injured individuals, and self-adjusting computer programs to monitor and advance the skill levels of users over time. With further increase in knowledge, new systems will be able to accommodate input from whatever interface mechanism the child is best capable of using, and to provide output using the sensory modalities which are most effective for any particular individual.

At present, progress in the areas of facilitating therapy and education is modest in comparison to the eventual potential. Although computer programs that can provide stimulation to the young and exercise individuals' skills do exist, they usually possess only a single function, and require the continual presence of trained personnel. This is especially true when current systems are used with severely retarded persons or with individuals who have short attention spans or low motivation. Also, these computer programs are rarely variable enough to maintain the attention of more severely handicapped children over long periods of time.

Despite the fact that technology is still new and software is not yet available for most therapy needs, these are not the primary barrier to rapid expansion of technology in this area. Rather, it is our limited ability to understand how the new capabilities can be effectively incorporated into the therapy and skill development processes. Incorporating the new tools into these processes requires a significant reorientation of our previous practices. Most therapists learned their techniques and procedures through a long process that includes extensive observations and practica in clinics and schools. In-depth educational mechanisms such as these do not yet exist for the advanced technologies, and will not be established for some time.

Another factor which may slow the development of applications of technology

in this area is the constraint which results from existing third-party reimbursement procedures. For example, will costs associated with therapy conducted without a therapist present be reimbursable?

Unfortunately, until these questions of funding and the training of personnel in the application of these technologies are resolved, the amount of money invested in the development of more effective software and hardware therapy tools will be limited. This dilemma will be ameliorated somewhat by federal grant funding processes that address the problem through grant support for the development of model programs and practices. Still, it is likely that it will be some time before the full potential of technology in therapy and training settings will be realized.

Provision, Restoration, or Extension of an Individual's Physical Abilities

The second application of technology in the rehabilitation process is to increase directly a client's capabilities. While the focus in therapy and education described above is on the temporary use of technology to improve an individual's skills, technology can also serve as a prosthetic or orthotic device designed to provide (or restore) a function or ability which the individual has lost.

The most successful application of technology to restore function has been with individuals who have either physical or sensory limitations. For individuals who are unable to manipulate, write, draw, see, hear, or speak, the new technologies are providing alternative mechanisms to meet their needs. This is done either by amplifying their existing abilities, or by transforming their interactions with the environment so that they can use an existing ability to compensate for an ability that is impaired.

For example, a speech-to-text translator might transform spoken English into a visual form for deaf people. A print-to-speech or print-to-Braille

device could transform printed text into either voice or Braille in order to better match the tactile and auditory abilities of a blind person. Special communication aids that are optimised for the residual motor movements of severely physically involved individuals can provide them with the opportunity to transform these limited movements into letters, words, and sentences. The output of these devices can be either spoken (through a speech synthesizer), written (to match the requirements of their education and work environment), or fed into computers as "keystrokes." In this fashion, individuals who cannot speak or write or use standard computers can have their present physical capabilities transformed to allow them to effectively converse, write, and access standard computers using standard software.

Some applications in this area can be very simple, as in the modification of electric battery-operated toys to allow severely disabled children to play (Wright, 1985). Others are more advanced, as in the modification of appliances and computers to permit disabled children and adults to operate them. Children who had previously been unable to draw a straight line can now draw perfect lines, circles, and scenes, using computers with special interfaces (Goldenberg, 1979). Interfaces are now available which can be operated by erratic control of the hand, movement of a muscle, or even eye gaze. In one case, a special "keyboard" has been developed, and is commercially available, which can be operated by simply looking at the "keys" (Friedman et al., 1984). For younger children, pictures or arrows can be placed on the "keys" to allow the children to manipulate objects on the computer screen or to draw pictures. Older children can make use of pictures and letters on the screen to provide them with a mechanism for communication and writing.

For older or more fully developed people, computer systems can be used to manipulate objects on a screen in order to develop special concepts, and to carry out activities (such as chemistry experiments) which they would otherwise

be unable to do, due to their severe physical involvement. In addition, individuals with disabilities can be enabled to carry out activities requiring precision, and to create final products (assignments, papers, etc.) without the smudges, smears, or distortions that were previously unavoidable due to their physical limitations. The effect of possessing these abilities on the self-esteem and motivation of persons with disabilities cannot be overestimated.

> A young boy with cerebral palsy is unable to speak, write, or use a keyboard. Using a portable "lap" computer which sits on his wheelchair laptray, the boy is able to use his head to control a spot on the screen of the computer. Using this spot, he is able to select letters, words and sentences to communicate. He can send his output to a voice synthesizer for conversation, to a printer for written work, tests, and assignments, or into the computer's memory for recall at a later time. He can also use his computer with its headpointing program to control the other computers at school that he must use for his physics and math classes.
>
> Mary has a similar system that she controls with her eyes.

Thus, new technologies can not only be instrumental in providing individuals with disabilities with the capabilities required for their development and education, but can also provide them with the experience of success and accomplishment that is important to their development.

Of the three general roles of computer technology in rehabilitation described in this paper, this second area is the most advanced, although it is still at an early stage in its development. Greater flexibility and portability of computer technologies will lead to more effective applications in this area. The major barrier to progress, as we have emphasized earlier, is the scarcity of persons trained in the selection, adaptation, and fitting of such aids. A secondary problem is the current state of third-party funding processes, which is limited to therapy and medical intervention in the rehabilitation and restoration of function for disabled individuals (and usually does not include

the provision of assistive devices as a third option).

Opportunities for Greater Participation in Regular Activities

The third role of technology in the rehabilitation process is less obvious than the first two, and is often overlooked. As our society becomes increasingly technology-based, it has the potential for becoming more accessible to individuals with a wide range of residual physical abilities. Unfortunately, it also has, at the same time, the potential for becoming less accessible, and for posing significant new barriers to disabled individuals (Vanderheiden, 1983).

As more electronically controlled devices appear, and as more jobs involve electronically controlled information processing and other procedures, the possibility for access to those jobs by individuals with physical and sensory disabilities increases. Devices that are controlled electronically lend themselves to alternate control mechanisms much more easily than devices which must be manipulated directly. Televisions, radios, appliances, doors, and even window blinds are now available that can be controlled electronically. Such systems are easily adapted for control by special electronic control systems designed specifically for people with physical or sensory disabilities.

The development of new technologies also facilitates the participation by disabled individuals in educational and employment settings. People who may have had difficulty in using books or printed materials because of visual, physical, or learning disabilities will find that information presented in electronic form is much easier to "manipulate." It is also easier to transform it into spoken or other forms that are more easily processed. Similarly, as computer simulation is used increasingly in physics, chemistry and other science courses, there will be a greater opportunity for physically disabled individuals to participate side by side with non-disabled students. For example, chemistry

glassware and acids, which could not be manipulated by an individual with a motor disability in a regular science classroom, will be easily manipulated using specialized "keyboards" as these experiments are implemented on electronic computer systems. Similarly, it will be possible for disabled individuals, using such systems, to conduct genetics experiments, physics demonstrations, and even to run a nuclear reactor, through simulations on a computer.

> Jimmy has severe athetoid cerebral palsy and an aptitude for science. Early on, he became fascinated with chemistry and would spend hours with his sister watching her work with her chemistry set. Ordinarily, chemistry -- which involves the use of a lot of glassware and acids -- would be out of the question for someone with severe athetoid cerebral palsy. His school, however, had just purchased a Chemistry Lab Simulation series of computer programs. (It was a small school, and couldn't afford a real, fully equipped laboratory -- so they were adapting the simulation approach earlier than most schools). Fortunately, the simulation programs were for the Apple computer, for which Jimmy had adaptations. He is now able to "handle" and "manipulate" laboratory equipment via the simulations on the computer display screen. With this necessary "hands-on" experience, he now plans to go on for a chemical engineering degree.

The trend toward electronic simulations in education is assured, because they will provide much wider experiences for all students, and at lower costs. Similarly, although the increasing use of new technology in offices and advanced educational programs is not deliberately <u>aimed</u> at benefitting the disabled individual, it can, nonetheless, do much to benefit him. In the coming years, there will be more educational, recreational, and vocational settings which are potentially more accessible and usable by people with a wide variety of handicapping conditions. This expectation, of course, is based on the assumption that these electronic information, control, and vocational systems will be constructed in such a way that the alternate input and output mechnisms needed by disabled individuals can be used with them. If these systems do not allow the use of alternate input and output mechanisms, then, in fact, they may pose new barriers rather than removing them.

Concerns and Reservations

The great potential of these technologies will not be realized without further efforts followed by careful implementation. There are many places where the advancing technologies can in fact impair and impede disabled individuals, rather than advance their cause. The inappropriate application of technology and the rapid incorporation of technology in our society have the potential to leave individuals with disabilities further behind at the end of this decade than they are at the present time. Some areas for concern are:

1) provision of tools without development of skills to use them;

2) premature application of new technology;

3) lack of access to the electronic society of tomorrow.

Provision of Tools Without Skills

All too often, technical aids are provided to individuals with the expectation that the mere provision of the device will enable the individual to use it effectively. It is important to keep in mind that these new technologies and aids are simply tools (Rodgers, 1984). The tools will be useless unless the persons using them are provided with the skills to use them correctly. It is not uncommon to see a person removed from a communication development program after he has been provided with a communication aid because "he now has a mechanism for communicating." In fact, he now has only the physical means with which to communicate, and is more in need of a communication development program than he was without the device. It is most important, also, that clinicians and teachers receive regular, formal training in order for them to understand what devices are available and how they may best be used. They must also be given the techniques necessary to help their clients effectively incorporate these tools into their skill development programs. Signs that tools, but not

training, have been provided are:

a) special aids and equipment that sit around, unused, in classrooms, rehabilitation programs, or closets.

b) children who have specialized aids but do not use them spontaneously or effectively.

Unfortunately, these are still common and familiar situations. It is often easier to secure funding to acquire new hardware for our programs than it is to secure funding for staff training that would enable them to use and incorporate these same technologies effectively in intervention programs.

Premature Application

A second area of concern is the premature application of technologies. Often, in an attempt to move a child on to more advanced forms of communication, a more advanced communication aid is provided to the child, despite the fact that he/she has not mastered the simpler forms of communication yet. In fact, in some cases, a child who is not making any progress on a simple communication system is provided with a more advanced system, under the belief that advanced technologies imbue advanced skills or hasten the development of advanced skills in their users. This often leads the intervention personnel and others in the child's environment to expect performance far above the child's current level, causing them to set intervention and therapy at a level which is inappropriate for the child. The most common result is a concerted effort to teach the child how to operate the aid, rather than to teach the child the basic communication or interaction skills which he or she lacks and needs help in developing.

This is not to imply that advanced technologies are not appropriate for young or retarded people. In fact, it will probably take more advanced technologies to be able to intervene effectively with the more severely involved clients. For example, a communication aid with voice output may have much greater motivational value, and be much more obvious in its operation, to a

severely retarded individual than would be a passive communication board which provided no direct feedback to the user. However, neither kind of communication board would be of much value to a child who is not already spontaneously communicating in some way, e.g., via gestures, facial expression, eye contact.

One of the dangers in the premature application of technology, then, is that people provided with a technology which is above their level will be expected to operate at that level, with therapy and intervention adjusted to these new "expectation" levels. Then, when the individuals prove unable to operate at that artificial level, they are all too apt to be downgraded in their "learning ability". The primary problem, of course, is not their lack of learning ability, but that the intervention and therapy they are receiving is beyond their level of understanding or development.

Lack of Access to the Electronic Society

Society is continually moving toward greater electronic integration. Soon, computers and information systems will be as integral a part of the home and the classroom as telephones, pencil, paper, and chalkboards are today. A Sears catalog can now be presented as pictures on a television screen, complete with "live" demonstrations of selected appliances. It is already possible to order catalog items by punching numbers on a keypad. Classrooms are using computers not only for presenting math, chemistry, and physics, but also for poetry and English lessons. It is estimated that before 1990, 75 per cent of all jobs will involve the use of computers in some way (Naisbitt, 1984).

If disabled people are able to use these computer-based systems, they will be able to participate in daily living, education, and employment opportunities to a much greater degree than ever before. If, however, these systems are not designed in such a way that they can be accessed by disabled people, these people will find increasing portions of society barred to them. For example, if

use of the information systems requires that the individual be able to use a keyboard, a pointing screen, or other input system which requires fine motor control, then many moderately to severely physically disabled individuals will be unable to use the systems. Similarly, systems which can be used only by those who are able to read CRT (TV) displays will not be accessible or usable by individuals who have severe visual impairments.

The situation can be compared to the situation which is currently occurring at many airports and other large buildings. In order to facilitate people moving about the airports, "moving sidewalks" are being installed. These sidewalks allow people to "walk" at twice the rate they would normally, thus allowing them to cover great distances more quickly. These systems, however, cannot be used by individuals in power wheelchairs, since the sidewalks are too narrow. Thus, everyone else is allowed to move more quickly than the disabled person.

The problem does not stop there, however. Once the moving sidewalk exists, the tendency is to create longer and longer hallways at the airports. Thus, the disabled individual is not only unable to access these accelerated transportation systems, but is also faced with the fact that the existence of these accelerated transportation system causes the design of buildings to change and increases the distances which must be travelled, increasing the handicap the disabled individual experiences. What will be the impact of moving sidewalk technologies on the lives of disabled individuals when these technologies spread from the airports to shopping centers, companies, and even the community? Similarly, as electronic technologies are incorporated into our society, it will not only make the society easier to access for those who are able to use these technologies as they are implemented and found in the public sector, but they will also cause the way in which we organize ourselves and do business to change, making it even more difficult for those individuals who cannot access

the standard computers, information systems, electronic library card catalogs, automated bank tellers, electronic directories, electronic catalog and ordering systems, and electronic education and employment settings.

Thus, it is important that steps be taken in the design of the equipment that drives our electronic society to ensure that access to the electronic information systems is provided. These access points could allow disabled individuals to connect their own special input or display devices to the information systems and to have the same access as their non-disabled peers.

Work is being initiated in this area. A special initiative from the Office of Special Education and Rehabilitative Services and the White House is bringing computer manufacturers together with researchers, consumers, and manufacturers of special adaptive interfaces to explore the best way to achieve this goal. The electronic information aspect of our society is developing very rapidly, however, and concerted efforts across a broad front will be required if we are to avoid the creating of an electronic information-based society that excludes the disabled.

Summary/Conclusions

It is clear that advancing technology is providing us with many new electronic tools, and that they are changing the shape of our society. These changes have the potential to affect disabled people of all ages either positively or negatively. For some applications such as automated therapy, the technologies may have a depersonalizing effect if not carefully applied. In other applications, technologies may provide means for conversation, mobility, and participation that greatly increase the disabled person's ability to interact with others. The effectiveness of these technologies in helping the disabled will depend on both our general knowledge and the skill level of the

people who introduce the technology to disabled persons and on those who help them in applying it. The new technological devices are only tools, which must be applied within carefully designed intervention programs. They will also be useful to the disabled individual only to the degree as they are appropriate to his or her level of skill and understanding.

We must move early and effectively to guarantee that access to common computers is provided for the highly specialized interface devices that may be used by disabled people. If, however, we are not able to develop or cause such access points to be installed, the electronic information systems will simply advance the productivity of everyone in society except those with disabilities, and thus leave persons with disabilities with a net loss in terms of their ability to compete and participate in our society. Furthermore, this loss is likely to be severe, particularly for the more able of the disabled population.

Implications for UAFs and Title V Programs

The expanding technology briefly described above has direct implications both for the operations of UAFs and the policies and planning activities of Title V programs.

UAF Considerations

As has been described elsewhere, the major responsibilities of a UAF include the training of health care personnel in the provision of services to people with a chronic handicapping condition; the application of newly acquired knowledge in the development and/or modification of service delivery programs; and the dissemination of that knowledge into community-based programs. New technology that can be used to intervene successfully in the debilitating effects of a handicapping condition should be included in each of these activities. It is important that trainees, service delivery personnel,

personnel from support organizations and educational programs, the families of clients, and clients themselves be aware of the opportunities available to them.

Contribute to expanding knowledge base. Perhaps the major aspect of a UAF that most distinguishes it from other clinical-based settings for the care of persons with a chronic handicapping condition is the emphasis placed on the academic nature of the program. The UAF emphasizes the development of new knowledge. In the past, particularly in the case of physical disabilities, UAFs have emphasized either rehabilitation as a correcting measure, or, in some cases, corrective surgery. The new technology discussed in this chapter suggests that a third approach is sometimes available -- the restoration of function or the provision of a substitute system capable of performing certain functions. UAF programs are ideal settings in which to develop those alternatives, to apply that technology when appropriate, and to examine the effects of the inclusion of the technology in long-term treatment or rehabilitation plans.

Interaction of engineers and clinicians. Another characteristic of UAFs that is important to the application of this new technology to the treatment of the disabled is that they are excellent settings for genuine interdisciplinary efforts. In this case, success is dependent upon the close interaction of specialized engineering personnel who design and construct the new biotechnology with the clinicians responsible for the care and treatment of disabled people. Just as the implementation of an artificial hip depends upon the productive interaction of an engineer and a surgeon, the development of a new communications system for a person with a communication-related disorder depends upon the productive interaction of a communications engineer and behavioral scientists skilled in speech/language processes, audiology, and occupational and physical therapy.

Appropriate setting for development and delivery of new technology services. The development of successful clinical services related to this new technology requires two critical elements: a relatively specialized facility dedicated to the development of the technology and a clientele that will be available for extended periods of time and which will be willing to participate in attempts at modifying and perfecting the new technology. At least initially, the kinds of services discussed in this paper will not be available in all communities within a region, because of limited numbers of personnel trained in such services and limited resources. Hence, initial efforts must be centralized in a few regionalized health-care facilities that already possess a broad range of disciplines. The thirty-six UAFs and eight satellites, each serving a relatively stable clientele within their regions of responsibility, are ideal for such developments.

Need for dissemination. No matter how effectively new technologies are developed, their potential contribution is limited by the extent to which the new capability is disseminated to programs and services outside the university environs. To the extent that UAF personnel work with Title V organizations, and other agencies and organizations who share in the care and treatment of disabled persons, the UAFs must play an important role in disseminating this new knowledge.

New training programs to provide personnel capable of introducing new technology. Finally, the most important role of a UAF is to prepare health care professionals who will promote and provide health care services of the highest quality. These services must include, in addition to more traditional expertise, knowledge about and provision of services involving "intervention" using new technological devices.

Title V Considerations

In addition to the important role of UAFs in the development of technology-

based services for persons with a chronic handicapping condition, it is important that a role be played by the Title V programs in each state. Although Title V programs have, historically, served as the focal point of the care and treatment efforts for children with a chronic handicapping condition, the developing new technologies provide new opportunities for Title V efforts. Thoese same new opportunities, however, also bring about new responsibilities. Some of the reponsibilities these programs might assume include:

Incorporate information about new services and programs into referral and information data bases. Recently there has been a great deal of discussion about regionalization of services for persons with handicapping conditions, including regionalized data base and referral systems and coordinated information base systems. Title V programs in each state are now responsible for assuring specified services for persons with a handicapping condition and for providing the resources to pay for those services in some cases. As new technology is developed, and as those developments translate into new opportunities for amelioration of handicapping conditions, it will be important to incorporate information about these opportunities into referral and information data base systems. Through these systems, the clinicians and residents of each state will be informed about the new opportunities.

Contribute to discussions about eventual establishment of standards for technology services. The development of new technological solutions to handicapping conditions will necessitate that we establish standards addresssing such issues as materials, protocols, and documentation procedures. There are three aspects to the issue of the establishment of standards. First, we must ensure the use of high-quality materials and procedures in the provision of services. Second, the rights and privileges of the client must be protected. Third, the appropriateness of new, technology-based services and their

eligibility for funding by public support must be documented. This third issue raises another difficult issue, but one which must be addressed: how to distribute scarce resources in an equitable way.

Incorporate considerations of technological intervention into long-range planning activities and in policy development. It is important to consider new development in health care and technology in planning and implementing programs that will address such issues as regionalization of services, establishment of coordinated state-wide programs, and development of resources for services. An example of a current key issue is the development of Diagnostic Related Grouping (DRGs) and their eventual impact on the provision of services to persons with handicapping conditions, especially those who might benefit by the application of new technology. How, for example, can clinical facilities resolve the conflict between, on the one hand, the desire to select materials and devices of the highest quality and provide intensive follow-up training and, on the other hand, the recognition that financial support for that care will be based solely on the diagnostic category in which the service falls. This issue, and many like it, must be anticipated, planned for, and eventually resolved. The solutions must be flexible enough to accomodate the continuing development of new technologies and their application to the fullest benefit of the client.

In summary, new technology is being developed that can provide significant new opportunities for more normalized functioning to persons with a chronic handicapping condition. That new technology, which involves both new equipment and new software, can be used to restore, facilitate, or replace limited abilities or functions. The success in applying the new technology, through new services for persons with disabilities, is dependent upon careful planning and implementation processes. Now is the time for those processes to be initiated. It is important for UAFs and Title V programs to lead those efforts.

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COMMUNITY BASED SERVICES FOR DISABLED/CHRONICALLY ILL CHILDREN AND THEIR FAMILIES

Merle G. McPherson, M.D.
Division of Maternal and Child Health
Bureau of Health Care Delivery and Assistance
Rockville, Maryland

The Philosophy of Community-Based Services

The concept of community-based services is far more than a current fad in health care. It is the logical 50-year development of, and state-of-the-art in, today's health care delivery system. The year 1985 marks the golden anniversary of the passage of the Social Security Act. In his January 4, 1935 message to Congress, President Franklin D. Roosevelt said: "Among our objectives I place the security of the men, women, and children of the Nation first." This statement, made three months before the passage of the Social Security Act, constitutes a declaration that the welfare of children was on a policy basis of equal importance with the welfare of adults. The Presidential Committee on Economic Security, in its report to the executive--a report which enumerated the various parts of the plan eventually to become the Social Security Act--emphasized that "the core of any social plan must be the child" and "every proposition we make must adhere to this core." This same report maintained that "Health measures that protect (the) family from sickness and remove the menacing apprehension of debt...are child-welfare measures." In addition, the Committee recognized the need for specifics not provided for in the general measures. The Crippled Children's Services (CCS), a principal focus of Title V of the Social Security Act (SSA), was originally directed to children with orthopedic handicaps, but Title V programs have extended their concerns to physically disabled, sensory impaired, developmentally disabled, and chronically ill children. Along with the extension of populations covered has been a

concomitant effort to devise a comprehensive approach which is child- and family-centered, rather than centered on the management of a specific problem or disease. This comprehensive approach, 50 years in its development, is the evolution of community-based services which allows children to move from institutions to the least restrictive, most psychologically appropriate setting for their development--that is, residing in their own homes and cared for in their own communities. As the President's Committee stated 50 years ago, the core of any social approach is the child, and the goal of community-based services is to recognize the child as a local citizen with the rights and responsibilities of living, playing, and growing in his own community. When we restore the child to his or her own family, school, and community, he or she actively participates in the intellectual, social, and recreational activities of his local peers, and receives health care from his or her own physician. Such a goal of community-based services requires changes in the service-delivery system to facilitate new arrangements with local physicians, school staff, community health, mental health, and social service providers--all of whom are working together to provide comprehensive and coordinated services. To be effective in both care and cost, these services must be:

1) Family- and child-centered in approach;

2) Developmental in focus;

3) Interdisciplinary in scope;

4) Individualized, active, and ongoing in nature;

5) Least restrictive in environment;

6) Comprehensive, continuous, and coordinated in execution.

Within these six standards, we make fundamental assumptions. As the child moves from the institutional to the home setting, the family shares in finding ways to provide and pay for care. Therefore, within the community-based

system, the State Crippled Children's (CC) agencies have as a given obligation the assistance to the family to become skilled caregivers and financial managers. Furthermore, when we focus upon the continuing development of the child from infancy through childhood, we must also analyze the interaction of the handicapping condition (be it orthopedic, sensory, respiratory, or cognitive) and the child's psychosocial development. If a service system considers only the "condition," the child will be a perennial patient.

The service system must be individualized, interdisciplinary, and continually monitored. These concepts inexorably lead to services which are both state coordinated and community based.

Regionalization

Inherent in the state-coordinated and community-based services system is the necessity for regionalization in order to provide the components of such a health system. Regionalization is the structuring of health services and resources within a given area to promote efficiency, avoid duplication, improve access, and assure quality. The historical development antedates the Social Security Act of 1935. The Armed Services had medical triage as the organizing principle of first aid, stabilization, and life support care. Through clinics, hospitals, and nursing-home care units in seven regions, the Veterans Administration has regionalized a national system of emergency, ambulatory, and inpatient care for millions of veterans. It is a system of sharing of equipment and services coupled with patient transfer for maximum use of resources. The Indian Health Service and the U.S. Public Health Service hospitals deployed resources in a similar manner for the Native American and Merchant Seamen populations, respectively. In a time closer to our own, the early 1950s, the Children's Bureau responded to the request of the American Academy of Pediatrics (AAP) for a regional system to deal with heart disease in children by funding Regional Congenital Heart Disease Centers providing both tertiary services to a

multi-state area and preparation of pediatric and surgical staff to perform the newly developing surgical techniques to repair heart lesions. This response to felt need added preservice and continuing education to the evolving regionalization philosophy. The following decades witnessed perhaps the greatest recent activity in Maternal and Child Health (MCH) regionalization in perinatal services. The 1960-1980 knowledge explosion in perinatal physiology and pathophysiology brought about the rapid expansion first of neonatal and then, perinatal, regionalization. Private professional organizations, often under public auspices, have also been responsible for standards for care and for regionalizing that care.

What history and practice have taught us is that successful application of regionalization is dependent upon the establishment of a strong community-based care system. While a tertiary care center stands at the hub of the system in its research, its exemplary interdisciplinary services and development of care plans, and its continuing-, preservice-, and inservice-education, it is ultimately the local physician and the community who use the research, implement the care plans, employ the education in day-to-day care management and habilitative services, and provide appropriate community support services to the family.

The decades of the 50s and 60s produced a twofold expansion in the development of community-based services as a component of Crippled Children's Services (CCS) programs. First, the emphasis broadened from the focus on the crippled child to the child with developmental, behavioral, and/or psychosocial problems. The Children's Bureau had been concerned with the problems of mentally retarded individuals even from its inception in 1912, but in the 1950s new directions emerged in approaches to services for mentally retarded children. Several disparate groups of parents joined together to form the National

Association for Retarded Children in 1950. Second, and as a corollary to the broadening of focus, came the recognition of the importance of community-based services. In a major address to the AAP in 1953, Dr. Grover P. Powers emphasized the expansion of medical responsibility and scientific concern to the psychosocial problems ensuing from the problems of the specific ailment. He encouraged the pediatrician to develop skills "...whereby he may help parents in their hour of black despair and offer them requisite time and encouragement to talk over implications and potentialities and possibilities, and also give them insight into community resources which he has perchance helped to develop."

Mental Retardation (MR) became a Title V program priority in 1954 with the publication of Report to Congress on the Conditions of Mothers and Children in this Nation. Within the Department of Health, Education and Welfare, the Secretary's Committee on MR was established to develop and support specific program components within various constituent agencies of the Department. The Title V agency was responsible for developing clinical services providing evaluation, diagnosis, and management assistance to families of children with developmental delays and/or mental retardation. By 1955, the Bureau had made several grants for special demonstrations of clinical services for mentally retarded persons. These clinics established a new focus--that of pediatrics--in their approach of pediatric direction, management responsibility, and a multidisciplinary team. Before these demonstration projects, medical direction was under a psychiatrist; the new direction marked the recognition of the need to look at the "whole child." It was also recognized that mental retardation was a broad problem beyond the management ability of a single discipline and more appropriate to a multi-disciplinary team who had skills beyond the expertise of the primary physician and who could coordinate the skills of professionals already in the MCH program delivery system.

The 1962 President's Panel on Mental Retardation presented over 90 summary

recommendations, including the need to develop a primary prevention program, the recognition of newborn screening and management of metabolic disorders as important steps in prevention, the support and development of cytogenetics laboratories, and the importance of the Crippled Children's program in its ability to provide corrective care for the secondary handicaps associated with MR. Not every problem of MR was solved. There were failures in the deinstitutionalization program and insufficient emphasis on State CC programs. Even though the advantages of the MR legislation of the 1960s outweigh the deficiencies, these inadequacies were undoubtedly responsible in part for the strong health mandate in P.L. 94-142 and the Developmental Disabilities legislation. Past errors underscore the need to develop to the utmost integrated community-based services to overcome duplications and deficiencies in federal legislation.

The decade of the 80s saw the Vanderbilt Study, "Public Policy Affecting Chronically Ill Children." This study delineates another, extended group of children requiring long-term management--those with rheumatic diseases, diabetes, hemophilia and other health impairments. One categorical disease, hemophilia, has further confirmed the need for strong community-based services, and the voluntary sector in conjunction with government groups has devised a model program with several aspects applicable to all those with specyal needs. For example, in the U.S., two-thirds of those with hemophilia receive care through a network of centers and satellites. The problems of the disease itself--low prevalence, need for high technology, and high cost of care--have brought about the development of a successful regionalized program contributing many facets to a community-based system:

- o home care as a prevention approach;
- o financial counsellors as members of the health care team;

- o development of psychosocial services as an integral component of comprehensive care;
- o patient and provider education as a component of comprehensive care
- o vocational and rehabilitative services; and
- o transition from child to adult care.

Both the rheumatic fever program and the hemophilia program have contributed many problem-solving devices and strategies, most of which are based on the community-based system as both ancillary and necessary to regionalization.

Collaborative and Networking Experiences in the Further Development of Community-Based Services

The Division of Maternal and Child Health and the Office of Special Education and Rehabilitative Services (OSERS) have since 1971 instituted and continued productive collaborative efforts to provide needed services to handicapped children and their families. These activities have allowed both agencies to work together at the state level to define community strategies and appropriate roles. The goals of this collaborative effort include:

- o improvement of identification of needs;
- o expansion of work with states to improve integrated service delivery at the local level;
- o provision of national leadership in defining "best practices" and in promulgating standards of care
- o improvement of the use of discretionary grant programs to facilitate and support efforts at state and local levels; and
- o improve coordination between private, public, and voluntary resources.

The projects outlined below are some of the efforts undertaken in this collaborative effort.

Improvement of Services Delivery at the Local Level: The State Collaborative Project under DMCH and SEP/OSERS

In 1977, these projects were created to demonstrate that agencies and professionals in health and education can work together to provide needed services to disabled children. The two agencies provided funds to Connecticut, Hawaii, Iowa, Louisiana, Oregon, and Utah to serve both preschool and school age populations, to identify barriers and constraints between health and education agencies, and to solve problems. In Connecticut, for example, a Medical Developmental Child Find was developed through public and private providers' identification of infants at risk, provided follow-up for this population, and developed curriculum for the professional training needed to put the model in place. In Utah, planners developed a project to transfer information from health agencies to school systems.

Throughout the six states, amelioration of services to children with special needs occurred. Interagency agreements of identification, evaluation, and service provision were developed. There was increased cooperation between health, education, and social service agencies and increased cooperation between agencies and individual professionals.

Public, Private, and Voluntary Resources: The American Academy of Pediatrics (AAP) Project

In 1982, sponsored by SEP/OSERS, the AAP and the National Association of State Directors of Special Education carried out a project aimed at state-level coordination to address the "related services" issue in P.L. 94-142. The section mandating the provision of "related services" to handicapping conditions specified neither which services were related nor what agency was to pay for

these services. In the six chosen states (California, Iowa, South Carolina, Colorado, Missouri, and New Jersey), a Statewide Advisory Task Force addressed the problems and provided solutions and better coordination of services. Each State Department of Education was given advice on improvement of services.

Improving Services for Preschool Handicapped: The Head Start Projects

In 1978, a Federal Interagency Task Force with representatives of the Division of Maternal and Child Health, Head Start, the National Institute of Mental Health, the Health Care Financing Administration, the Special Education Program, and the Administration on Developmental Disabilities supported the development of a project by Head Start in Washington and Oregon aimed at the 0-5 year age group. The American Association of University Affiliated Programs (AAUAP) under the leadership of Dr. Phyllis Magrab at the Georgetown University Child Development Center UAF provided consultation to this effort, which developed two manuals to facilitate the development of community-based systems of collaboration between service agencies and providers.

Assisting State and Local Communities in Improving Services to Children with Special Needs and their Families: The Georgetown Network Project

The Network Project offers ongoing consultation, technical assistance, and resource sharing for the representatives of the previously described Collaborative Projects, AAP projects, and Head Start projects. Representatives from these projects met at the Georgetown University Child Development Center UAF in 1983 to develop a network and a unique sharing, dissemination, and coordination of expertise on community collaboration to improve services,

standards of care, and financing of care for children with special needs. Technical assistance includes:

- o state planning for local involvement
- o local planning for state involvement
- o attitudinal training of community professionals
- o establishment of standards of care
- o establishment of systems of continuance of care management
- o development of creative financing strategies
- o development of management information systems
- o solutions for related services' issues
- o establishment of identification systems
- o consideration of special rural and urban planning and implementation issues

The Network Project unites health, mental health, education, and social services agencies in collaborative efforts. The methodology of this collaboration includes:

- o Direct linkages among states;
- o Development of materials (workbooks, resource files, etc.);
- o Brokering of resources across states; and
- o Individual consultation to states.

Improving Early Intervention Programs: Project Zero to Three

Because statutory authorities often overlap more than they dovetail, and because administrative arrangements vary considerably among jurisdictions, our efforts to develop integrated community based systems are sometimes difficult to accomplish, but necessary. For many years the State Maternal and Child Health and Crippled Children's Services programs have provided early intervention services for infants with discernible handicaps and their families.

More recently, other federally supported programs such as Developmental Disabilities, Early Periodic Screening Diagnosis and Treatment, Head Start, Education of the Handicapped (P.L. 94-142), Title XX, Social Services, and the National Institutes of Mental Health have also emphasized the importance of early intervention in the age group "Zero-to-Three." MCH concern for this age group is a part of the concept of a continuum of care starting at conception, proceeding through infancy, childhood, adolescence, and the childbearing years. MCH is facilitating the integration of efforts to improve services to infants and toddlers with handicapping conditions --Project Zero to Three. This project, designed to assist one state in each of the DHHS regions in the development of model programs to be extended through knowledge sharing, has a multifaceted programmatic structure. The project serves as a training program for health and education personnel, a dissemination program for sharing information and experience among all the states, a material-development program directed to current state needs, and a program for coordination of consultation services (as requested) to address specific state issues. The accomplishment of these goals is through meetings from representatives of the participating state programs, through technical assistance and consultation with staff of Project Zero to Three, regional offices, and participating states, and through consensus building. The UAF at the University of Iowa is currently preparing a "best practices" monograph "Early Services for Children with Special Needs: Transactions for Family Support."

All of these projects reflect contemporary developmental theories recognizing the importance of the biological and social interaction between the child and his or her environment. They are all multidisciplinary in scope. They all are dependent upon the complementary roles of public, private, and voluntary resources. They are all geared to a model of community-based services

to children with special needs.

State SPRANS Experience

Under the set aside section of the MCH Services Block Grant (Title V, Social Security Act, as amended) the Secretary of DHHS is authorized to provide for Special Projects of Regional and National Significance (SPRANS). A number of states are developing important demonstration programs supported by the SPRANS grants. Of the several which could be cited, the following five offer a representative sample.

The Massachusetts Tri-Agency Project for the Development of Policy and Program Strategies for Handicapped Children and Their Families is a program devised by three organizations (Division of Family Health Services [DFHS], DPH, the Developmental Evaluation Clinic UAF in the Children's Hospital, and the Department of MCH and Aging in the Harvard School of Public Health) to design and implement a state-wide comprehensive system of care for handicapped and chronically ill children and their families. The goals at the initiation of Project SERVE were needs assessment and policy analysis and implementation of a comprehensive system through consultation and technical assistance to the DFHS. The most recent phase of this project has been the development of strategies for alternative models for financing and for health care delivery. Project SERVE was formulated as a review of the operation of the State Clinic System, and is to assist the DFHS define its present and future role in service delivery to this population.

The outcome of this project will be a state system of health care for this population. This system will be coordinated with other providers, public and private, and will provide an equitable and rational design for services. The project, a cooperative project of the three child health agencies enumerated above, focuses on improved integration of existing services and more efficient use of existing funding and will have relevant products and strategies

replicable in other regions.

Another project is the "Coordination of Care for Chronically Ill Children" conducted by Health Research, Inc. in Albany, New York and the New York State Department of Health. The general purpose of this program is coordinating care for specified chronic illnesses of children among medical entities and voluntary support organizations with specialized care teams. The program will establish demonstration projects as a coordinated, comprehensive service model, and will see to the coordination of plans in the department of health and other agencies. Importantly, it will establish a data system of numbers of children, age, condition, and location of care. Regional workshops throughout the state and feasibility studies on the nature and scope of the problem will be included as well. The program will demonstrate, on a pilot basis in selected regions, that the care of chronically ill children can be organized and coordinated in a way that provides for comprehensive, continuous, and cost-effective services. It is anticipated that the program will increase the quality of care at home and will have a positive effect on psycho-social counseling, subspecialty professional services, educational needs, and financial needs of this population. The second year continuation of this project includes a supplement to examine costs of care within this model.

One of Iowa's programs, "The Networking of Services for Handicapped Children," is a demonstration of the state government's compliance with Title V MCH Block Grant legislation. Programs formerly under the jurisdiction of the federal government are now state directed with appropriate input from the communities. Regional networks coordinate available services needed by chronically ill children and their families. The program has begun the initiation of a data/information system to facilitate interagency communication and evaluation of care delivery systems.

The project will increase knowledge concerning the needs for services and outcomes of services, will create a more effective method for provision of services through interorganizational cooperation, and will demonstrate a new model for collection and analysis of multiagency data used for evaluation and planning. The continuation of this grant will be spent in consolidating support services for chronically ill children, in a continuing education program for Network Integrated Evaluation and Planning Clinics, and in multiprofessional agency medical counseling and treatment services for adolescents. A study of methodology will be published at the end of Year 2 of the grant.

The next exemplary program cited is one devoted to a specific disease, but one with techniques transferable to other community-based programs--the Juvenile Diabetics Health Maintenance Program of the Arkansas Department of Human Services in Little Rock. This program has as its purpose the development of a state-wide system of care to improve the quality of services for children with Insulin-Dependent Diabetes Mellitus (IDDM). The goals of this program are to develop linkages between primary, second, and tertiary levels to assure coordinated and comprehensive care and to develop a state-wide education program on IDDM for professionals, supporting agencies, school personnel, and parents and families of those affected. In addition, the program plans a central registry for coordination of assistance in medical equipment and supplies, insurance, and rehabilitation. As well as providing mechanisms to help normalize blood sugar, plasma lipids, and hemoglobin A.C. values for young patients with IDDM, the program demonstrates that home monitoring will be effective both in treatment and in cost.

An important aspect of the community-based service delivery program is that it is interdisciplinary in scope. The final exemplary program cited, "Community Health Care for Children with Chronic Conditions," is an interdisciplinary, multifacility project to stimulate the development of comprehensive community-

based services for this population. Centered at the Gillette Children's Hospital UAF in St. Paul, Minnesota, the program offers a novel three emphases:

1) The development of a regional consortium composed of professional disciplines, institutions, and public and voluntary categorial disease agencies. The consortium is a study group for exploration and facilitation of cooperative efforts.
2) The core effort will be diversification of community-based activities by programs with expertise in chronic disease.
3) The analysis of current patterns of health-care financing.

Comprehensive health care for children and youth with chronic illnesses is a national priority. This project addresses effective regionalized systems of health care, community support systems, and improved methods of financing of health care.

Major Issues and Problems

Since the inception of the State CC programs in the 30s, there has been a broadening of scope of populations served and a change in perspective in the treatment offered. From a program geared to children with orthopedic handicaps, there has been an evolutionary expansion to children with other sensory disabilities and mental or motor problems. The definition of crippling conditions was expanded to include provision for children with congenital heart disease, cleft palate, amputation, retardation, and other handicaps. In the late 60s, Pediatric Pulmonary Centers were federally funded, and by the early 70s Sickle Cell screening, education, and counseling clinics were established under The National Sickle Cell Anemia Control Act. This legislation was superseded by the National Sickle Cell Anemia, Cooley's Anemia, Tay-Sachs and Genetic Disease Act of 1976 to cover all genetic diseases. The Comprehensive Hemophilia Diagnostic and Treatment Center Program came into existence through

Section 1131 of P.L. 94-63. At present in the U.S., the DMCH participates in providing services to the entire gamut of chronically ill and disabled children. As well as expansion of population treated, there has been a notable shift in focus from that of treatment which is disease centered to that of treatment which is child and family-centered. This expansion of population and mode of treatment has not occurred without concomitant problems which can be summarized briefly under three categories: the systemic constraints, the societal constraints, and the financial constraints.

The evolution which has occurred has been influenced by historical events. The systemic constraints arise from the differences in the public policy of the 60's--one based on extensive social legislation--and today's policies. The 60s approach often seemed to bypass state government, while the current federal trend has been to return responsibility to the states. There has been reduction of federal dollar support and a reduction of categorical legislation to block grants.

In the 1980s, State CC agencies have greater responsibility but less statutory and regulatory authority because of block grant legislation. The problem is compounded by the increasing number of children needing services and by the increasing costs of the medical care required by these children. Other systemic needs for change are brought about by biomedical and technological advance. Surgical advances in congenital heart disease, improved prosthetic and orthotic technology, blood concentrate therapy for hemophilia, and ventilation support for bronchopulmonary dysfunction are just some examples of the forces exerting pressure for developmental change. The system must fund the methods to encourage these new developments and to supply them to children in need of them. In addition there are societal problems associated with the advances. Legislation has been passed which includes Amendments to Title V of the Social Security Act, the provision of appropriate education to all handicapped children

(P.L. 94-142), the provision of advocates and appropriate service programs for persons with developmental disabilities (P.L. 94-103), and for the prevention of discrimination against persons with handicapping conditions in federally funded programs (P.L. 95-602). These legislative mandates make civil rights for handicapped individuals of paramount concern in the agenda of public and private agencies. From provision of care to children with designated crippling conditions to provision of care for children in a comprehensive systematic health care program, regardless of handicapping condition, there have been significant pressures for change. It has been pointed out (in The Unexpected Minority--Handicapped Children in America) that Americans with handicaps often are targets for unconscious social and political oppression, "often more damaging to their lives than their physical disabilities." Ironically, society's view of the handicapped as perennial patients is humane in intent but society's perception of the "handicap" role may cause harmful psychological and sociological consequences through society's expectation that someone with a handicapping condition should always remain in the role of a "good patient."

Professional realization that members in this population have developmental characteristics similar or identical to so-called "normal" children has brought about the recognition that it is neither necessary nor desirable to segregate this population from the rest of society. This has led to the trend that moves away from keeping a chronically ill child in an institutional setting and toward creating a less restrictive environment by caring for him within his own community.

The major economic constraint is obviously the increasing demand for services coupled with the decreasing supply of funds for chronically ill children. Accelerated cost rates for public and private third party payors has shifted downward the level of state CCS agencies' support for chronically ill

children. Quality of care maintained under cost-effectiveness is the criterion by which programs must now be judged.

Since all of these political, social, and economic concerns interact, the need to organize providers and recipients into a service delivery system is obvious. We must devise a systematic, systemic approach in response to the biomedical, political, social and economic forces and constraints in order to provide needed services to chronically ill children and their families. This interactive relationship among the forces and constraints therefore leads to a systems approach in service delivery--an approach based on the view of the child as a member of a microsystem, the family, which in turn is a part of a larger system, the community. A group of communities constitutes a state-based system, economically and societally responsible for providing regional service delivery.

Future Efforts

In addition to projects described in this monograph, a central issue is the future directions of services to be provided by the federally funded (Title V MCH Block Grant) CCS programs. The National MCH Resource Center of the University of Iowa has a project to examine the following issues:

- o an historical overview of governmentally supported programs for children with special health care needs;
- o the services needed and the best methods to provide them;
- o the barriers to obtaining these services;
- o the organization of governmentally supported programs providing services at the state and local level with emphasis upon the creation of community-based networks of integrated services; and
- o the financing of these services.

A national report will be published to enunciate goals and objectives and to furnish guidance to state CC administrators, to state and local public policy makers, and to other organizations, groups, and individuals responsible for, or

interested in, the delivery of services to these children. This project and its multifaceted report will deal with the major problems of the state CC programs and other public programs, as well as the fragmentation of services for handicapped and chronically ill children, the problems of casefinding and location, inadequate reporting systems, and funding of health related services in an economy of inflation. The preliminary studies made by this project indicate a reflective and innovative approach, and the initial outline of the study indicates a thorough investigation of the topic. The final report should be a landmark document.

As is obvious throughout this report (e.g., the Georgetown University "Network Project," the University of Iowa UAF "0 - 3 Project," and Project SERVE), UAFs are involved in many of the programs currently funded through Title V programs. The relationship of the UAFs and Title V should continue to expand. For the UAF, the regional mode based on community-based services is a natural one since their structure allows them to extend beyond single local governments. The UAF, by virtue of its academic orientation and its familiarity with new technology, serves as a rich resource to work closely with the state CC program in the development of community-based programs--the evolutionary development of fifty years of Title V. The new directions of Title V are thus determined by opportunities in a pluralistic system,--a strengthened State Title V system, the SPRANS grants including UAFs and other systems--which will maximize the effective use of all available resources--public, private, and voluntary.

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THE FRAGILE-X SYNDROME

David L. Meryash, M.D.

Child Development Center (UAF)
Rhode Island Hospital
Brown Program in Medicine
Providence, Rhode Island

The fragile-X syndrome is an inherited genetic condition associated with developmental disability which was recognized as a distinct entity only during the past decade. It can affect several members of both sexes in a single family, although it predominantly affects males. The fragile-X syndrome is a relatively frequent cause of mental retardation occurring among all major races and cultures in the United States, and is now estimated to affect one out of every 2000 males. It is important for practicing physicians, special education teachers, public health planners, and other professionals who provide care to individuals with developmental disabilities to be familiar with the fragile-X syndrome.

The name of the condition refers to the peculiar appearance of the X chromosome demonstrated on chromosome analysis of affected persons.

Historical Aspects

For many years prior to the identification of the fragile-X syndrome, it had been noted that there existed an excess of males among populations of mentally retarded individuals. Reports of families in which there were large numbers of mentally retarded males were reported in the literature during the 1940's and 1960's. These included those described by Martin and Bell (1943), Allan, Herndon, and Dudley (1944), Renpenning (1962), and Dunn et al. (1963). These families all suggested a pattern of X-linked inheritance. Lehrke (1972) at the University of Wisconsin hypothesized that there were one or several

genes on the X chromosome that lead to mental retardation. Several X-linked entities -- Hunter syndrome and Lesch-Nyhan syndrome, for example -- were distinguished by the early 1970's because they are associated with specific enzymatic deficiencies. There remained a large number of families with apparent X-linked mental retardation in which affected individuals had no distinguishing features. Collectively these cases were known either as nonspecific mental retardation or as Renpenning syndrome.

In 1969 Lubs reported a family in which two mentally retarded brothers demonstrated a secondary constriction at the distal end of the long arm of their X chromosomes. At the time his paper received little notice. In the mid-1970's there emerged reports of families with apparent X-linked mental retardation in which affected males had macroorchidism. These individuals were reported as being cytogenetically normal. At the same time Sutherland in Australia was studying the occurrence of fragile sites, or a tendency in some families for chromosomes to break at specific points. Some of the families which he studied were families in which there was X-linked mental retardation. He discovered that the mentally retarded males in those families consistently demonstrated fragility at the distal end of the X chromosome. What soon became apparent was that, in order to demonstrate the fragile site on the X, the medium used to culture the cells had to be deficient in certain nutrients, specifically folic acid and thymidine. In the interval between Lubs' report in 1969 and the examination of families with X-linked mental retardation in the mid-1970's, the culture medium generally in use had been improved to include all kinds of nutrients to enhance cell growth. What Sutherland noted was that this added nourishment masked the expression of fragile sites in general and, in particular, the one on the X. He also noted that those males who demonstrated the fragile X chromosome had macroorchidism. Turner, Daniel, and Frost (1980) and others confirmed that individuals with X-linked mental retardation

and macroorchidism had the fragile X chromosome. Thus the genetic world recognized the "macroorchidism marker X (MOMX)" or the "fragile-X" syndrome. The families described by Martin and Bell and by Dunn et al. turned out to have the fragile-X syndrome, while those presented by Renpenning and by Allan, Herndon, and Dudley did not.

Hundreds of families with the fragile-X syndrome have now been reported in the medical literature. It is now confirmed that the fragile-X syndrome is, after Down syndrome, the second most common identifiable cause of mental retardation. It has attracted the attention of medical geneticists throughout the world perhaps because it is not only a fairly frequently occurring condition, but because it is one of the few truly major discoveries in the area of mental retardation to come along in many years. The interest of geneticists and developmentalists has been maintained also because the fragile-X syndrome has proven to be a challenging disorder to study. Over the past five years perhaps more questions have been raised than answered.

The Fragile-X Chromosome

The diagnosis of the fragile-X syndrome is confirmed in a mentally retarded male by the observation on chromosome analysis of a fragile site on the X. The tip of the long arm of the X chromosome appears loose and either attached by only a narrow strand or completely detached. The site at which the separation occurs (Xq27) is consistent from cell to cell and from individual to individual. It is not clear what the properties of the chromosome are at that particular site which lead to breakage, but it is known that demonstration of the fragile site in the laboratory depends on culture conditions which deplete the pool of thymidine available for DNA synthesis. A deficiency of folic acid is one such condition. In affected males the fragile X chromosome can be

demonstrated in from several to 50% of the cells examined, but not all. The frequency with which the fragile X is demonstrated bears no relationship to the severity of symptoms in a particular male patient.

Turner et al. (1980) were the first to report that the fragile X chromosome could be demonstrated in some mildly mentally retarded females. These affected women and girls were, according to Turner, phenotypically normal. Family studies identified other mentally retarded relatives including both males and females who demonstrated the fragile X. It has since been shown that roughly one out of three women who are carriers (or heterozygotes) for the fragile X condition demonstrate fragility at Xq27 on one of their two X's.

Physical Aspects/Health Issues

Males with the fragile-X syndrome have mild to severe mental retardation and frequently have characteristic physical and behavioral features. They have relative macrocephaly, dolicocephaly (head length proportionately greater than head breadth), long, narrow faces, and prominent ears. The majority have short stature (Meryash et al. 1984). Males with the fragile-X also have prominent jaws. Some investigators have described this as "prognathism," which means that the mandible and the lower teeth protrude beyond the maxilla and upper teeth. Careful dental studies on a group of individuals have determined that actual "prognathism" is not a feature (Lindner, 1985). An early impression was that affected males had large hands and feet. The reverse was shown through anthropometric studies (Meryash et al. 1984). The initial reports also suggested a tendency toward light irides, a characteristic now thought to have been the result of ascertainment bias. For an as yet undetermined reason, males who have reached puberty have larger-than-normal sized testes. Large testicular size has occasionally led to the diagnosis in a developmentally delayed infant (Carmi et al. 1985); this characteristic may even be present

prenatally (Jenkins et al. 1984). No major medical problems are known to be specifically associated with the fragile-X syndrome. The enlargement of the testes is unassociated with endocrinologic abnormality and, as far as is known, does not interfere with reproductive capacity. There may also be a somewhat increased frequency of hypospadias and cryptorchidism among males with the fragile X (Meryash, Hazen, & Gerald, 1983).

Although some affected individuals have seizures, the prevalence of seizure disorders is not greater in this group than among individuals with mental retardation from other causes.

The enlarged head size is probably related to a combination of megalencephaly (large brain) and thickened calvarium. Although CAT scans will often show increased ventricular size, hydrocephalus is not a feature, the large ventricular size being more likely the result of underdevelopment of brain. This is a nonspecific finding among individuals with mental retardation from a variety of developmental causes.

This investigator has noted no specific neurological abnormalities among individuals with the fragile-X syndrome. However, young children with the disorder usually are somewhat hypotonic, not unlike children with other forms of mild to moderate global developmental delay.

Recently some investigators (Pyeritz et al. 1982; Hagerman et al. 1984) have noted ligamentous laxity and mitral valve prolapse in some of the individuals with the fragile-X syndrome they have examined, and they suggest that these findings are indicative of an underlying connective tissue disorder. These features have not as yet been found with any great frequency among people with the fragile-X syndrome examined by other investigators. Although two individuals described by Hagerman had aortic dilatation, the health-related importance of mitral valve prolapse alone is not clear since it is present in

about 4 to 6% of the general population.

Two men with the fragile-X syndrome who lived in the Boston area have had postmortem examinations. Both died in their thirties, one of aspiration leading to respiratory and cardiac arrest, and the other of a massive myocardial infarction. The first had been healthy in life, but had fibrotic tissue in his heart on autopsy; the second in life had poorly controlled hypertension. Neuropathologic findings in both patients were nonspecific.

Developmental and Behavioral Aspects

Although the degree of mental retardation covers a wide range, the majority of males with the fragile-X syndrome are mildly to moderately retarded. In general, children who are living at home and enrolled in special education programs tend to be higher functioning than older men who were institutionalized many years ago, or for whom special education was not available when they were young children. Men and boys with the fragile-X syndrome are usually socially engaging; however, they commonly have an unusual style of interacting with other people. For example, they tend to avoid direct eye contact during conversation; that is, they have a gaze aversive tendency (Rose et al. 1983). Their speech characteristically is rapid, dysfluent, agrammatical, and has atypical prosody (intonation patterns). Young children are often characterizable as hyperactive. Older children and adults, although they can sit in one place for extended periods, are usually excessively anxious and "fidgety."

Although cognition is globally depressed, males with the fragile X perform particularly poorly on tasks involving short-term memory (Rose et al. 1983).

Some males with the fragile X (fewer than 5%) have classic infantile autism -- they have nearly no useful speech and lack responsiveness to other people (Meryash, Szymanski, & Gerald, 1982; Brown et al. 1982). However, the

majority of affected males have "autistic-like" behaviors, for example gaze aversiveness and stereotypic movements (repetitive combing of the hair or rocking); thus, a higher than 5% frequency of autism is reported by some investigators who use a less-stringent definition.

There have been a few scattered reports of intellectually "normal" males who demonstrate the fragile X chromosome. This is an apparently rare occurrence since the general population surveys which have been done to date failed to identify any such individuals. The men who have been reported also did not have adequate psychological testing to absolutely rule out subtle cognitive impairment. More troubling, as discussed below, is the suggestion that some males may carry the gene or genes that transmit the fragile-X syndrome yet neither be retarded nor demonstrate the fragile X chomosome.

When cognitive deficits are present in female carriers, they are usually milder than those seen in the affected males. However, some affected females do have severe mental retardation and physically resemble the mentally retarded males.

Treatment

Education and Other Therapies

Treatment for an individual with the fragile-X syndrome currently consists of an individualized program of special education including such components as speech therapy, physical therapy, and vocational counseling as necessary. An affected child less than three years of age can benefit from participation in an early intervention program. Children with the fragile-X syndrome are eligible for public support of these programs through P.L. 94-142.

Medical Treatment

As yet there is no proven medical treatment specific for the fragile-X

syndrome. A few affected children who are "hyperactive" and have attentional difficulties have been helped with medication (e.g., Ritalin) which is used for children of normal intelligence who have similar problems. Folic acid, a vitamin, is currently being evaluated as one possible way to help individuals with the fragile-X syndrome (Brown et al. 1984; Hagerman et al. 1983). The fact that folic acid in the laboratory masks the fragility of the X chromosome has led some researchers to test the efficacy of the vitamin for treating individuals with the condition. Results so far of this experimental treatment have been inconclusive at best. Folic acid levels and folate metabolism so far have been shown to be normal in individuals with the fragile-X syndrome (Erbe, 1984). Moreover, it is not even known what the relationship is between the fragility of the chromosome and the occurrence of developmental disability. The chromosomal breakage may actually only be a laboratory phenomenon. Certainly further study in this area is necessary.

Genetic Aspects/Genetic Counseling

The fragile-X syndrome generally follows a pattern of X-linked inheritance. Two important characteristics of X-linked inheritance are satisfied: predominance in males and no father to son transmission. Recent studies by Sherman et al. (1984), however, suggest that genetic transmission of the fragile-X syndrome may be more complicated.

X-linked conditions occur as the result of an altered gene on the X chromosome. The differences in frequency and severity between the two sexes can be explained by the fact that males have only one X chromosome, but females have two. If one of a woman's two X chromosomes carries the gene for a particular disorder, the effects of this gene can be compensated for by the corresponding "normal" gene on her other X. A male's Y chromosome does not

have the same genes as the X. If a male's X chromosome, then, has the abnormal gene, he will usually have the characteristics of the disorder, because there is no "normal" gene to mask the effects of the abnormal gene. In each of a female's cells, one of her two X's is randomly inactivated. The variation in expression of a particular disorder in females can be explained by the proportion of normal X's versus abnormal X's which are inactivated. In the fragile-X syndrome this mechanism, known as the Lyon hypothesis, can explain why some female carriers are retarded while the majority have normal intelligence. In the fragile-X syndrome, however, females are more often affected than in other known X-linked disorders.

A child inherits one sex chromosome from each parent -- one of the mother's two "X" chromosomes, and either father's "X" or father's "Y" chromosome. If the child inherits father's "X", the child is a girl -- if the child inherits father's "Y", the child is a boy.

If a woman has the fragile-X gene on one of her X chromosomes, she is heterozygous for the fragile-X gene and a "carrier" for the fragile-X syndrome. The daughter of a carrier has a one out of two (50%) chance herself of being a carrier. A carrier's son has a one out of two (50%) chance of having the fragile-X gene.

A man who has the fragile-X gene gives each of his daughters his only X chromosome. Thus, <u>each</u> of his daughters will be a carrier. Each of the sons of a man with the fragile-X gene receives his "Y" chromosome, not his "X". Thus, none of his sons will have the fragile-X gene.

The study by Sherman et al. (1984), a computer-assisted analysis of 110 pedigrees of families with the fragile-X syndrome, found a statistical deficiency of affected males. The investigators concluded after studying their data that this meant that there were silent male carriers as well as silent female carriers. Specifically, they cautioned that as many as one out of every

five males who carry the gene for the fragile X neither demonstrates the fragile X chromosome nor is mentally retarded. The only way these men could be identified as carriers, therefore, would be if they had affected daughters or affected grandchildren of either sex. If this is the case, genetic counseling for the fragile-X syndrome becomes very complicated indeed. Proposed mechanisms for this apparent nonpenetrance in males include the involvement of more than a single gene in the fragile-X story.

Determination of Carrier Status

A woman who has had more than one son with the fragile-X syndrome, or who has one son and an affected father or brother, is an obligatory carrier. Similarly, each daughter of an affected male is an obligatory carrier. Thus, one can often determine if a woman is a carrier simply from her family history.

Approximately one out of three female carriers will demonstrate the fragile site on one of her two X chromosomes. Thus, a woman from a family in which the fragile-X syndrome occurs is identified as a carrier if she demonstrates the fragile X chromosome. Mentally retarded carriers often convincingly demonstrate the chromosome change. However, only about one out of three carriers is mentally retarded or has significant learning disability. Carriers with normal intelligence either demonstrate the fragile X chromosome in only a small percentage of cells or not at all. It is estimated that one out of every 1000 women is a carrier for the fragile-X syndrome. Thus, there is currently no reliable laboratory test to detect the majority of women who are carriers.

For any female relative of an affected male who is not an obligatory carrier and who does not show a fragile X chromosome, a risk for her being a carrier can be calculated based on how she is related and on whether she has already

had any normal sons and how many. The method for doing this, called Bayesian analysis, is one of the tools of genetic counselors and medical geneticists.

Several laboratories have been trying different methods for enhancing the expression of the fragile site in carrier females, but demonstration of the fragile X in non-retarded carriers remains difficult. Researchers in other laboratories nationwide are studying more modern methods (use of recombinant DNA technology and "restriction fragment length polymorphisms" or RFLPs) for the identification of carriers and non-carriers in already identified families.

Prenatal Diagnosis

There is presently no completely reliable method for prenatal diagnosis of the fragile-X syndrome except to determine the sex of the unborn child. The fragile X chromosome has been demonstrated in cells obtained during amniocentesis from women who are known carriers as well as by sampling of fetal blood cells obtained by fetoscopy from the umbilical cord (Brown et al. 1984; Shapiro et al. 1982). However, the future development of the child who, as a fetus, either demonstrates a fragile X chromosome or does not have a fragile X chromosome cannot be predicted with certainty at this time. Improved methods for diagnosing the fragile-X syndrome during early pregnancy will also, like detection of carriers, depend on recombinant DNA technology.

Establishing the Diagnosis and Follow-up

Mentally retarded or autistic children should be strongly suspected of having the fragile-X syndrome if:

1. One or several siblings are also similarly affected and/or the child's mother has a mentally retarded brother, or if

2. The child's mother is mildly mentally retarded.

The diagnosis is more likely if:

3. The head circumference of the retarded child is more than two standard deviations above the mean for his age, or his head circumference is proportionately large for his height, and

4. The child does not have severe neurologic impairment or major congenital malformations. The presence of either would suggest a diagnosis other than the fragile-X syndrome.

If the diagnosis is suspected, a chromosome analysis should be requested by the child's physician through a laboratory that has had considerable experience with the procedure for demonstrating the fragile X chromosome. Instructions should be given to the laboratory to look specifically for the fragile X, together with a clinical description of the patient and a family pedigree. If several members of a family are mentally retarded, it is best to request a chromosome analysis for affected males before females are studied because of the greater ease of demonstration of the chromosome abnormality in affected males.

Once the diagnosis is made, the family can be referred to a genetic center for counseling. The geneticist or genetic counselor will assist in determining which members of the family should have their chromosomes studied and in determining the risk of various relatives for having affected children.

Additional Counseling Issues

Parents of a child or children diagnosed as having the fragile-X syndrome occasionally ask why, despite careful examination, the diagnosis was not made previously. The phenomenon in which mental retardation affects several males within a family has been known for many years. However, it was not until the late 1970's that the fragile X chromosome was found repeatedly among individuals in these families, and it was recognized that special conditions in

the laboratory were required to demonstrate the chromosome change. If a child's chromosomes were examined prior to 1980 or so, it is unlikely that the techniques used would have demonstrated the fragile X chromosome.

It should be kept in mind also that the fragile-X syndrome is not the only form of X-linked mental retardation. Families are often encountered in which there are two or more mentally retarded males who either fit the description of another "syndrome" or who still cannot be given a diagnosis. The affected males in Renpenning syndrome, for example, have microcephaly (as opposed to large head size), normal-sized testes, severe mental retardation, and do not demonstrate a fragile-X chromosome.

If a single child in a family with no other history of mental retardation is diagnosed as having the fragile-X syndrome, the family may well wonder how this could have occurred if the condition is hereditary. There are several possible explanations:

1. As mentioned above, the mother of an affected boy usually has normal intelligence. Each of her brothers has a 50% chance of being affected and a 50% chance of being unaffected. The larger her family, the more likely she will have an affected brother, but if her family is small, or if she has no brothers, there may no family history of mental retardation that she is aware of. If she inherited the trait from her mother, who also had a small family, the condition may be absent from several generations.

2. We are not certain of the range of disability among males with the fragile-X syndrome. It is possible that many affected males in past generations have functioned adequately in their lifestyles and occupations, so that they were not recognized as having any difficulties.

3. A third possibility is that the child is affected as a result of a new mutation. Sherman et al.'s study (1984) suggests that mutations leading to affected males are rare if they occur at all. This would imply that the mother

of a child with the fragile-X syndrome is a carrier even if only a single son is affected.

Because the fragile-X syndrome is a relatively common condition, many centers in the United States and around the world are engaged in research into improving diagnosis, possible treatment, and prevention of the disorder. It is particularly important to find out more about the nature of the handicaps experienced by individuals with the fragile-X syndrome and the way they learn, so that the best educational strategies can be used with young children.

Implications for Title V Programs and UAFs

University Affiliated Facilities (UAFs)

Despite modern biotechnology and increased professional and public attention to the area of developmental disabilities, the underlying cause still cannot be determined for nearly one out of three mentally retarded children thoroughly evaluated in child development clinics. The recognition in the past decade of the fragile-X syndrome as a distinct entity that can be confirmed by laboratory examination has contributed to reducing the number of children with mental retardation for whom the cause must remain unknown. Moreover, awareness of the fragile-X syndrome provides not only an explanation of causation to many parents of children with mental retardation, but also a means for estimating recurrence risk for these parents and for other relatives.

As facilities with responsibility for identifying children with handicapping conditions, for characterizing those disabilities, and for training professionals in various disciplines in the evaluation and care of children with developmental disabilities, UAFs have a major role in the fragile-X syndrome scenario. Mentally retarded children evaluated by UAFs who fit the description presented above should be examined cytogenetically for the fragile-X

chromosome. When a child is diagnosed as having the fragile-X syndrome, the physician should work to identify others in the family who may be affected as well as those who are at-risk for transmitting the condition. He or she should make sure that the family receives genetic counseling. The other professionals on the evaluation team should assist with educational planning and psychosocial support. Each UAF should ensure that the genetic laboratory with which it is affiliated is well-trained in the technique of examining for the fragile X chromosome abnormality and is kept up-to-date on new developments regarding diagnosis. UAFs should be supportive of research involving developmental and behavioral aspects of the syndrome. They should also encourage the Mental Retardation Research Centers to make a priority the development of more accurate methods for carrier detection and prenatal diagnosis, specifically through the use of genetic probes. Lastly, UAFs should keep informed of new discoveries and recommendations regarding the diagnosis and treatment of the fragile-X syndrome so that they can disseminate this information to community agencies and facilities that care for the developmentally disabled.

Maternal and Infant Care Programs and Family Planning Programs

One of the important phenomena associated with the fragile-X syndrome is the frequent occurrence of intellectually limited mothers. It is especially important that these mothers be identified and given relevant supports inasmuch as they can have genetically normal children as well as mentally handicapped children. Women who have the fragile X gene who are themselves mildly mentally retarded are "at-risk" for having difficulties with daily living skills including parenting and should be counseled with special sensitivity. They should be offered family planning alternatives as would intellectually normal carriers.

Genetic Disease Programs

Based on the estimated prevalence of the fragile-X syndrome amoung live-born males (1 out of 2000), many families remain unidentified. Because of the wide variation of the cognitive deficit, it is likely that many "affected" individuals function adequately enough within their everyday environments so as not to be identified as "abnormal." This is very apt to be the case in non-urban communities. State-run genetic programs which provide regional services locally are in the best position to identify many such families who have the fragile-X syndrome. Establishment of the diagnosis in these more remote family units can lead to identification of individuals in the extended family who are at-risk for transmitting the condition. Regionalized genetic programs can facilitate the counseling of individuals who would never receive services if they had to travel to a major urban center.

Thus, collectively UAFs and Title V Programs can act to:

1. Identify individuals with the fragile-X syndrome and facilitate their receiving relevant services, including genetic counseling.
2. Educate professionals, the public, and governmental health-planning agencies about the fragile-X syndrome.
3. Provide psychosocial support to affected families.
4. Support research regarding developmental, behavioral, and physical aspects of the fragile-X syndrome.
5. Encourage research into improving carrier detection and prenatal diagnostic methods.

TABLE

CLINICAL FEATURES OF THE FRAGILE-X SYNDROME

PHYSICAL FEATURES

Short stature
Absolute or relative macrocephaly
Dolicocephaly
Long-narrow face
Prominent lateral palatine ridges
Prominent ears
Prominent chin
Macroorchidism in males
Small hands and feet *
Ligamentous laxity *
Mitral valve prolapse *

* needs further study to confirm

DEVELOPMENTAL AND BEHAVIORAL CHARACTERISTICS

Hypotonia in infancy and early childhood
Global developmental delay
Hyperactivity during preschool years
Increased anxiety and fidgety behavior in later childhood and adulthood
Gaze aversiveness
Dysfluent speech
Agrammatism
Atypical prosody
Stereotypic behaviors
Classic infantile autism (occasionally)

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II.
A CONSIDERATION OF LINKAGES

Linkages Among UAFs and State Title V Agencies

Elaine M. Eklund, M.S.
AAUAP MCH Consortium Project

Of the many areas in which the University Affiliated Facilities (UAFs) and State Title V programs may collaborate, it is the training area that provides the most natural bridge for cooperation. With training as the foundation of their mission, the UAFs represent unique resources for the delivery of training for clinical skills in dealing with the disabled and chronically ill populations served by the state programs. Those agencies, in turn, provide to the UAFs a source of clients for a clinically based training program and represent a major system which will eventually employ UAF trained personnel.

In practice, training may be the single focus of working arrangements between the UAF and State Title V program or it may be but one component of a multi-faceted arrangement. In the following pages, examples of a variety of agreements, with training as the major focus, are described. As noted, the training does not always occur in isolation and the examples frequently illustrate the blending that occurs with other components such as service delivery, shared staffing arrangements, advisory relationships, or mutual research endeavors.

The descriptions of linkages or working arrangements are not all inclusive but are examples from which we may learn. Many are merely the current steps of multi-year activities which have also been explored in earlier editions of this publication.

The chart on the following pages depicts visually the points of interface in working relationships over the past year. Reports were obtained utilizing the UAFs as the point of contact for developing the information in cooperation with personnel of the various state Title V programs with which they worked.

Table 1. UAF and MCH/CC Linkages 1985

Degree of Involvement
1 - Periodic
2 - Moderate
3 - Substantial
* - New or Emerging

	Center for Developmental & Learning Disorders Birmingham, AL	Division of Developmental Disabilities & Clinical Genetics Irvine, CA	University Affiliated Facility Mental Retardation Program Los Angeles, CA	University Affiliated Training Program, Children's Hospital of LA Los Angeles, CA	Georgetown University Child Development Center Washington, DC	Mailman Center for Child Development Miami, FL	Riley Child Development Program Indianapolis, IN
Training							
Shared Trainees	3	1		*	1		1
Workshops/Seminars	3	2	2	2	2	3	3
Inservice Provided through Outreach	3	*	2	2	1	2	2
Extended Inservice at UAF Site	3	*	2	3	1		
Courses/Lectures	3	2		2	2	3	2
Trainee Practicum or Workstudy		3		2		3	1
Trainee Experiences in Clinics or Services	1	3	3	3	2	3	2
Service							
Operating MCH or CC Specialty Clinics or Services in UAF	3	3		*		3	
UAF Staffing of MCH- or CC-Operated Clinics	2	1	3	2			3
Follow-Up	3	3		*	2	3	3
Outreach to Provide Service to MCH or CC Clients	3		3	2		2	*
Service Provided on Contractual Basis	2	3		*	2	3	*
Providing Service on Referral or Cross-Referral Basis	3	3	3	3	2	3	3
Advisory							
Special Topic Advisory Committee, e.g., Genetics, Developing Standards	1	3		3	3	3	3
Title V State Administering Agency		3		1	3	3	
Crippled Children's Program	2	2	*	3		3	1
MCH Program	1	3	*	3	3	3	1
SPRANS Program		3		3	3	3	1
Title V Personnel on UAF Advisory Committee	1	*		1			2
Interagency Committee for Communication or Liaison	2		2	3	2	3	
Staff							
Employment of UAF Trainees in Title V Programs	3	3	1	2	1	2	2
Shared or Jointly Funded Staff	1			3		2	2
Professional Consultation	2		3	3		3	3
Planning							
Developing Processes or Systems, e.g., Case Management, Needs Assessment	1	3	3	3		3	1
Regional Collaborative Projects	3		1			3	2
Research							
Utilizing Health Records	1	2	1	2	1		1
Shared Research			2	2		3	3

Division of Developmental Disabilities Iowa City, IA	Kansas University Affiliated Facility-Kansas City Kansas City, KS	John F. Kennedy Institute for Handicapped Children Baltimore, MD	Developmental Evaluation Clinic Boston, MA	Eunice Kennedy Shriver Center for Mental Retardation Waltham, MA	Developmental Disabilities Program St. Paul, MN	University Affiliated Facility for Developmental Disabilities Kansas City, MO	Meyer Children's Rehabilitatin Institute Omaha, NE	University Affiliated Facility Piscataway, NJ	University Affiliated Facility Rose F. Kennedy Center Bronx, NY	University Affiliated Program for Developmental Disabilities Rochester, NY	Mental Retardation Institute Valhalla, NY	Division for Disorders of Development & Learning Chapel Hill, NC	University Affiliated Cincinnati Center for Developmental Disorders Cincinnati, OH	The Nisonger Center Columbus, OH
	1	1		3		2		1			2	1		
1	2	2	2	3	2		1		1		1	3	3	2
1	2	3	2	3	2	2	3		2		2	3	2	2
	1	1		1							1	3		2
2	2	1	2	3	2	3	1				2	3	2	2
2	1	1		2		3					*			2
2	3	3	3	3	2	3	3	1	3	3	3	2	3	2
	1	3	3		2		2	3	3	3	3		3	
1	3	1	3		2		3	2			1		*	1
3	3	1	2	2	2		2	2	3		2	3	3	2
1	3	3	3	1	2			2	3		3	3	3	
1	3	3	2		3	1	3	3		3	2	3	3	
3	3	3	2	2	2		2	2	2		1	2	3	3
2	2	2	3	3	1	2	2	2	2		2	2	3	3
	1	2	2	1			2					2	3	2
1	1	2	3		1	1					1	1	2	2
1	1	1	2	1								1	2	
3	3	1	2	3	3						3		3	1
	1	3	*	3		1	1				*		3	1
	3	2	1	3		1					1	3	3	3
	1	2	2	3				1	3		2	2	2	2
1	2	2		-			2				2	1	2	
2	2	2	3	3	1	2	2		3		2	3	3	3
	2	2	2	2		1	2	2	3		2	1	3	2
2	2	*	3	3	1		2		3	3	2	1	3	1
1	2		3	1				*			1		1	1
1	1	3	2	3		1	1		*		*	1	2	1

Table 1. UAF and MCH/CC Linkages 1985

Continued

	Child Development & Rehabilitation Center Portland, OR	UAF Program of South Carolina-University of South Carolina Columbia, SC	UAF Program of South Carolina-Winthrop College Rock Hill, SC	Child Development Center Memphis, TN	University Affiliated Center Dallas, TX	Developmental Center for Handicapped Persons Logan, UT	Child Development & Mental Retardation Center Seattle, WA	Waisman Center for Mental Retardation & Human Development Madison, WI
Training								
Shared Trainees	3			2				1
Workshops/Seminars	3			3	3		2	1
Inservice Provided through Outreach	3			2	3		2	1
Extended Inservice at UAF Site		3	2	2	3			1
Courses/Lectures	2			3	3		2	
Trainee Practicum or Workstudy	1			1	3			
Trainee Experiences in Clinics or Services	3		1	3	2	1		3
Service								
Operating MCH or CC Specialty Clinics or Services in UAF				3		2	3	3
UAF Staffing of MCH- or CC-Operated Clinics	3	3		3		2		
Follow-Up	3	3		3			3	2
Outreach to Provide Service to MCH or CC Clients	3			1			3	3
Service Provided on Contractual Basis	2	3	*	3		2	3	3
Providing Service on Referral or Cross-Referral Basis	3		3	3	2		3	3
Advisory								
Special Topic Advisory Committee, e.g., Genetics, Developing Standards	1		1	3	2		2	3
Title V State Administering Agency				1	2			2
Crippled Children's Program	3			2	1			3
MCH Program				3	3			2
SPRANS Program	3			3	1		3	1
Title V Personnel on UAF Advisory Committee			2					3
Interagency Committee for Communication or Liaison	3			3	3	1		3
Staff								
Employment of UAF Trainees in Title V Programs				3			3	1
Shared or Jointly Funded Staff	3			2		2	2	1
Professional Consultation	3			3	3			
Planning								
Developing Processes or Systems, e.g., Case Management, Needs Assessment	3		1	3	3	2	3	3
Regional Collaborative Projects	3			2	3			2
Research								
Utilizing Health Records	3						1	2
Shared Research							1	1

Training Relationships

The range of ways in which training relationships are developed and implemented is almost as broad as the number of players involved. Conferences are traditionally, and continue to be, a primary mode for delivery of training. "Meeting the Nutritional Needs of Chronically Ill and Handicapped Children" was the subject of a conference developed for Title V personnel in federal MCH Regions IV and VI by the Center for Developmental and Learning Disorders UAF at the University of Alabama at Birmingham.

As a result of a Region III Nutrition Conference, planning for the coordination and expansion of nutrition services within the Washington, D.C. metropolitan area was initiated on a regular basis involving the District's MCH Nutritionist, the Director of Nutrition of the Georgetown University Child Development Center (GUCDC) UAF, and ten other nutritionists from public and private agencies. A year after the conference, the group had completed a needs assessment and developed a joint funding proposal for continuing education. In the same year, the CCS Nutritionist's position in the District of Columbia became vacant and a former trainee of the GUCDC UAF was selected for the job . Prior to starting work in the program, the CCS Nutritionist returned to the UAF for one week to update skills and, since then, has provided additional training for current GUCDC Nutrition trainees.

Collaborative training of students with a commitment to a career in MCH/CC is a joint effort now ready for implementation as a result of an agreement between the MCH Training Program in Handicapped Children of the San Diego State University's Graduate School of Public Health and the University Affiliated Training Program, Children's Hospital of Los Angeles. Teams of 3-5 trainees from the School of Public Health will enter a two year MPH program each year. During the first year they will be based in San Diego with selected activities at the UAF or with UAF faculty and trainees. During the second year, they will

be based at the UAF and will participate in selected activities with SPH faculty. In addition, the relationship includes linkages with two departments at the University of Southern California (Occupational Therapy and Physical Therapy) and with the Los Angeles CCS, all integral components to carrying out the plan.

In Tennessee the "Governor's Task Force on Healthy Children" has investigated efforts to develop a high risk registry. Staff of the Child Development Center (CDC) UAF have served on the Task Force and collaborated with state agency personnel on continuing education conferences, inservice education, and state wide planning for the registry. The CDC UAF Director has been appointed by the state MCH program to the advisory board for implementation of the high risk registry.

Planning was done for a jointly sponsored conference on technology dependent children between the Ohio Division of Maternal and Child Health and the Nisonger Center UAF. Nisonger cosponsored a conference on dental services with the Division of Dental Health.

The Nisonger Center UAF and University Affiliated Cincinnati Center for Developmental Disabilities UAF cooperated to provide a regional teleconference on "Ohio's Approach to Prevention." Audiences at eight sites in seven states and Washington, D.C. included personnel of MCH/CC Programs, Area Health Education Centers (AHECs), Developmental Disabilities Planning Councils, Cooperative Extension Services, and UAFs.

An agreement between the Indiana Division of Maternal and Child Health and the Riley Child Development Program UAF provides for reciprocal training opportunities. The Riley staff agreed to conduct a needs assessment and present a series of inservice programs on the medical/developmental needs of multiply handicapped children at the Silvercrest Children's Developmental Center, a state

operated short-term residential facility. In turn, the Silvercrest staff's expertise concerning issues of residential care of children is utilized in the UAF education and training curriculum.

A series of inservice training sessions for all social work staff of the Los Angeles County CCS was provided by the UCLA UAF both at the CCS sites and at the UAF site. The training focused on the use of psycho-social groups for working with physically handicapped adolescents. The sessions also included training experiences for UAF trainees and was provided through a service provision mode in which followup research was conducted.

Following up on a conference conducted in the previous year, the Georgetown University Child Development Center UAF (GUCDC) and the Southwest office of the Virginia Department of Health conducted a "Respite Care in Urgent Need" conference. The purpose was to develop an action plan for interagency collaboration necessary to meet the respite care needs of chronically ill children and their families. Invitees included representatives of health, education, social services and parents.

Also in a collaboration between the Virginia State Department of Health and the GUCDC, an effort to redefine and expand the role of the nurse within the State's Child Development Centers led to training provided first in the Northern Virginia area and later to be delivered to other areas of the State. Nursing staff also cooperated with the Virginia State School Nurse Achievement Program (SNAP) to provide inservice on sexuality in handicapped children and intermittent self-catheterization.

This year has produced increased training interaction between the Florida CC program and the Mailman Center UAF with the collaborative provision of formal continuing education programs for nurses, physicians, and other health professionals in a broad range of handicapping conditions and in child abuse and neglect. Topics in the series have ranged from the state of the art in prenatal

diagnosis to aspects of dealing with adolescents with chronic illness or disability.

Using a mix of Title V, state appropriations, and private foundation funding, an agreement between the Division of Developmental Disabilities UAF and the Iowa CC Program trains Pediatric Nurse Practioners for followup with high risk infants. At the request of the New York State Department of Health, a five day training program for public health nurses in developmental screening was developed by the nursing faculty of the Mental Retardation Institute UAF in Valhalla. Approximately 93 nurses from four counties were trained in administration of developmental screening for the Infant Health Assessment Program.

The Kansas University Affiliated Facility in Kansas City has been instrumental in coordinating continuing education activities for two projects in the Kansas Department of Health and Environment, Division of Health. The first was a workshop on Genetics for Health Professionals cooperatively planned and conducted by the KUAF, the Region VII Division of Health--Family Planning Training, and the Department of Pediatrics and Medicine of the Kansas University Medical Center. The second project, for identifying and tracking high risk infants, included developing a five state regional meeting to provide a forum for dissemination of information on issues related to high risk infants and to further develop regional networking among health, education and social service providers serving children birth to three. Other training has targeted community health nurses for training in genetics for health care professionals and multidisciplinary audiences for training in a systems approach to early childhood developmental services.

The most unique UAF-Title V linkage is in Portland, Oregon, where the Child Development and Rehabilitation Center UAF is a part of the Crippled Children's

Division. Although the extent of interrelatedness is not likely to be duplicated anywhere else in the country, many of the essential components that are shared here are also shared elsewhere. Moving outside both programs, UAF trainees receive "real life" training experiences within local public health offices.

In the past year, the Portland UAF has also participated with UAFs in Eugene, Oregon (Center on Human Development) and Seattle, Washington (Child Development and Mental Retardation Center) in a networking project for training in the states of federal MCH Region X. The project provides for ongoing assessment of training needs, identification of resources, and facilitation of appropriate interventions through continuing education, consultation and technical assistance activities. Title V program directors from Oregon, Washington, Alaska and Idaho serve on the Advisory Board.

Linkages for Service, Staffing, Needs Assessment and Advisory Relationships

As noted, linkages are not confined to training. Relationships built around providing service are numerous and mutually rewarding to the parties entering the agreement. Joint staffing agreements continue in a variety of forms. Title V staff serve on UAF advisory boards and UAF staff reciprocate on a wide variety of special topic advisory boards. The following is a sampling of the activity that has gone on in the past year.

The Southeast Asian Genetics Program (SEAGEP) is a program which illustrates the breadth of interactions which can be intertwined in cooperative activity. SEAGEP is an outreach program of the University of California, Irvine-University Affiliated Program for Southeast Asian refugees living in Orange County, California. The program includes documentation of the prevalence/incidence of genetic disease within the various Asian cultures, community and professional education, and screening for hemoglobinopathies. The UCI/UAP has been designated as an Area Screening Center for the new California Maternal Serum

Alphafetoprotein (MSAFP) Screening Program. A special advisory committee links the MSAFP Screening to the SEAGEP program. Linkages with the state Title V program are also evident in agreements for the state to work with the UAP for service provision and for training of state personnel by the UAP.

The Kansas University Affiliated Facility-Kansas City, is the recipient of a contract for a Kansas Crippled and Chronically Ill Children's Program (KCCICP) Coordinator to be based at the UAF and has a contract for nutritional services for KCCICP clinics.

At the Developmental Disabilities Program, Gillette Children's Hospital, (now a partner in the new Minnesota University Affiliated Facility Center) several arrangements for service provision are in effect with the Crippled Children's program. The CC orthopedic screening clinic is held at the UAF and is staffed by UAF physicians from pediatrics and orthopedics. The CC program and UAF also maintain an annual fee-for-service contract for eligible children and youth.

In New Jersey, several of the service programs administered by the University Affiliated Facility at the Rutgers Medical School are provided under contract with the state Maternal and Child Health Program, including programs of the Laurie Neurodevelopmental Institute and the Chronic Disease Program. These programs provide a range of services including diagnosis, treatment, follow-up, outreach, coordination and referral.

A staff pediatrician of the Division for Disorders of Development and Learning (DDDL) UAF in Chapel Hill has been charged with organizing a cerebral palsy clinic. She will work with staff of the Pediatric Rehabilitation Clinic in North Carolina Memorial Hospital and with DDDL staff to organize and staff the clinic. Across the State of Nebraska, MCH Genetics clinics are provided by the Meyer Children's Rehabilitation Institute.

In New York, the Rose F. Kennedy Center UAF has organized meetings leading to development of an interagency task force, including New York State MCH/CC staff, other state agencies, and representatives of the American Academy of Pediatrics to study and make recommendations about the "related services" issues involving children with disabilities in special education programs.

The Kennedy Institute UAF worked with the Maryland Crippled Children's Services to assess the nutrition services to children with developmental disabilities in Maryland. The project has involved identifying utilization patterns, barriers to service and location of available resources. An audit of CCS charts to suggest strategies for program planning, program management, and coordination of services was the final phase of the project.

In its second year, Project SERVE, a joint project of the Division of Family Health Services of the Massachusetts Department of Health, the Harvard School of Public Health, and the Developmental Evaluation Clinic UAF of the Children's Hospital, conducted a parent-based assessment of the needs of handicapped and chronically ill children, which obtained over 900 questionnaire responses.

Training Needs of State Maternal and Child Health and Crippled Children's Programs

Elaine M. Eklund, M.S.
AAUAP MCH Consortium Project

Background

In 1983, a survey of training needs of state MCH and CC Programs, designed and conducted by a subcommittee of the Association of MCH and CC Programs was undertaken in cooperation with the AAUAP's MCH Consortium Project. Responses were received from 44 programs in 33 states. Twelve of the responses were from CC programs, nineteen from Maternal and Child Health programs, twelve were from combined MCH and CC programs, and one was not identified. At that time, and still today, no comparable information existed that systematically catalogued the training needs of those programs. This article looks at the survey's methodology and the results that were obtained. The preceding paper, which reviews the linkages between University Affiliated Facilities (UAFs) and the State MCH and CC Programs, includes descriptions of working relationships in the training realm and, as such, is a logical followup to the training needs assessment. Indeed, over the past three years, the interaction between those programs has shown a healthy progression of growth which, in part, is a response to the needs described.

Methodology of the Survey

The survey instrument's first major focus was the identification of training needs--both preservice and inservice--in 14 topical service areas, by discipline, for 16 disciplines. Respondents were asked to indicate which of the training categories they felt should be required and which would be desirable

for each discipline. The training categories and disciplines are as follow:

Training Topics	Disciplines
Interdisciplinary Team Functioning	Administration
Family Support Services	Audiology
Genetics	Dentistry
Screening for Handicaps	Education
Developmental Testing/Diagnosis	Epidemiology
Case Management	Genetics
Behavioral Management	Nursing
Early Intervention	Nutrition
Fiscal Management	Obstetrics
Use of Statistics	Occupational Therapy
Evaluation of the Physically Handicapped Person	Orthopedics
	Pediatrics
Evaluation of the Mentally Retarded Person	Physical Therapy
Effect of Chronic Illness on the Family (Emotional-Social-Economic)	Psychology
	Social Services
Influences of Cultural Differences on Management of Health Problems	Speech

A second major area of inquiry was included relative to the diagnosis and management of specific conditions. The Title V Directors were asked to indicate their opinion as to the desirability of preservice training for personnel in six consolidated discipline groupings for the following conditions: Multi-Handicapped, Mental Retardation, Autism, Speeech Impairment, Hearing Impairment, Vision Impairment, Cerebral Palsy, Hemophilia, Genetic Disorders, Epilepsy, Sickle Cell Disease, Developmental Delay, Prematurity, High Risk Pregnancy (Medical Aspects, Social Aspects, Service Delivery, Prevention, and Screening).

Descriptive Information from the Reporting Programs

Respondents were also asked to provide minimal descriptive program information. Questions related to logistical and practical aspects of training, e.g., preference for training site, interest in staff exchanges, and budgeted funds available for training were included; opportunity for open ended comments or suggestions relative to training was provided as well.

Ninety percent of the responding programs indicated that they contracted for services. Of those, almost half (45%) contracted for more than 2/3 of their service program activities. Twenty-nine percent (29%) contracted for 1/3 to 2/3 of service program activities and 26% contracted for 1/3 or less.

In describing their programs' staff by discipline, respondents did not report the number of staff, but instead merely checked the disciplines listed. All disciplines were checked by one or more programs. The discipline most frequently reported among salaried staff was nursing, followed by pediatrics, social work, and nutrition (in order of frequency).

Although few programs were recruiting at the time of the survey, all disciplines except orthopedics were cited as ones that the programs would like to recruit if funds were available. The most commonly desired disciplines for recruiting (if funds were available) were nursing, social work, pediatrics, physical therapy and nutrition.

Results of the Survey

The analysis of the results was reported in two ways:

1) The ratio, or relative weight, of each training area, for example "Interdisciplinary Team Functioning" compared to "Family Support Services." (The relative weighting was calculated by dividing the total number of responses made for any training area by the total possible responses that could be made for that area).

2) The response to each cell of the matrix (expressed as a percentage), e.g., the percentage of respondents that perceive a need for pediatricians to be trained in "Use of Statistics."

Preservice Training Needs

Respondents' opinions were considerably stronger regarding what should be "required" preservice topics than with those considered "desirable." For instance, the topic ranked highest among those that should be required, "Interdisciplinary Team Functioning," received a weighting of .4950 compared to a weighting of only .3099 for the first ranked "desirable" topic, "Use of Statistics." The relative weighting of the latter is comparable only to the fifth ranked topic in the "required" list (Evaluation of Physically Handicapped). The top five weighted topics respondents felt should be required are shown in Table 1.

TABLE 1

"REQUIRED" PRESERVICE TRAINING

Topics in Rank Order	Weighting
1) Interdisciplinary Team Functioning	.4950
2) Case Management	.3572
3) Family Support Services	.3185
4) Early Intervention	.3013
5) Evaluation of Physically Handicapped	.2999

There was also greater spread of weights within these categories than with those identified as desirable, particularly between the first and second above (14 percentage points). The significance of that difference is not completely understood. One interpretation lies in the nature of the topics, i.e., inasmuch as the interdisciplinary approach is a delivery mode for the services (case management, family support, etc.), its relatively stronger weighting may reflect

a recognition of its fundamental importance to carrying out the agencies' varying priorities.

The five topics identified as most desirable for preservice training were:

1) Use of Statistics
2) Genetics
3) Influence of Cultural Differences on Management of Health Problems
4) Early Intervention
5) Family Support Services

The following table includes the relative weighting of each topic and the responses for each cell in the matrix.

TABLE 2

AREAS CONSIDERED MOST DESIRABLE FOR PRESERVICE TRAINING

Responses Reported as Percentages of Total Possible Response by Discipline

	Pediatrics	Nursing	Social Services	Nutrition	Orthopedics	Obstetrics	Genetics	Speech	Audiology	Psychology	Occupational Therapy	Physical Therapy	Dentistry	Education	Administration	Epidemiology	Rehab Weight of Evidence	Training Areas	
Use of Statistics	56%	54	44	42	27	42	37	32	32	17	27	24	34	39	17	5			.3099
Genetics	32	56	44	44	34	25	7	37	32	27	27	27	25	37	27	24			.2984
Influences of Cultural Differences on Mngmt of Health Problems	49	42	27	37	22	34	32	24	22	20	24	24	29	29	27	17			.2740
Early Intervention	29	27	40	37	24	20	22	37	39	20	24	27	24	39	32	17			.2697
Family Support Services	39	27	7	37	32	27	24	32	27	22	27	27	34	17	34	17			.2539

Inservice Training Needs

The program directors were asked to indicate areas of need for inservice training for both salaried and consultant staff and to respond in terms of current needs and in contemplation of future needs. Perhaps reflecting the immediacy of need, few programs responded to the question in terms of future inservice training needs, e.g., the greatest response to a single item was from 11 programs, of the 42 responding to the questionnaire, projecting a need for training in behavioral management for nursing personnel.

Current (in 1983) inservice training needs reflected the top ranked area as "Interdisciplinary Team Fuctioning", which was thus ranked for both salaried and consultant staff. The second highest ranked area was "Early Intervention" followed by "Genetics."

As with responses regarding preservice training, the greatest indication of need for professional groups was for pediatricians, nurses and social workers. In only three other areas did responses exceed 33% (34% indicating a need for nutritionists to be trained in Interdisciplinary Team Functioning and 40% indicating a need for administrative personnel to receive inservice training in the area of Fiscal Management and in the Use of Statistics).

The percentage of responses to each cell for nine training areas and the relative weights of those training areas are shown in Table 3.

TABLE 3

CURRENT INSERVICE TRAINING NEEDS

Responses Reported as Percentages of Total Possible Responses by Discipline

	Pediatrics	Nursing	Social Services	Nutrition	Orthopedics	Obstetrics	Genetics	Speech	Audiology	Psychology	Occupational Therapy	Physical Therapy	Dentistry	Education	Administration	Epidemiology	Relative Weight	Training Areas
Interdisciplinary Team Functioning	34%	51	51	34	20	20	20	20	20	17	14	17	20	17	26	9	.2336	
Early Intervention	29	31	31	20	14	14	11	17	20	14	11	14	9	17	6	6	.1580	
Genetics	34	46	26	17	11	17	20	11	11	9	9	11	11	11	9	6	.1563	
Developmental Testing/Diagnosis	40	49	20	11	9	6	11	17	14	14	9	11	6	14	11	6	.1479	
Case Management	23	46	34	26	9	9	11	14	11	9	6	9	9	9	17	0	.1429	
Effect of chronic illness on the family (Emotional-Social-Economic)	29	43	29	20	9	6	9	11	9	11	14	14	9	14	6	3	.1395	
Influences of cultural differences on management of health problems	23	34	29	23	6	9	6	14	11	14	11	11	6	11	11	6	.1345	
Screening for Handicaps	23	40	14	11	11	6	17	17	20	11	6	9	11	9	11	6	.1328	
Family Support Services	11	46	46	9	3	9	17	5	5	9	9	9	3	9	9	0	.1176	

The training areas listed in Table 4 are listed in rank order according to the relative weight obtained as a function of all responses. Interestingly, the second ranked of those, "Early intervention" received that ranking with a balanced need for training of personnel across the board without an extremely high percentage shown for any single discipline.

Other Areas of Inquiry

A few questions were included relative to logistical and practical aspects of training. For example, respondents were asked if they felt there were an advantage to a particular site for conducting inservice training. Eighteen said they preferred their own central office site, three suggested the UAF clinical setting, and nine indicated a preference for their own clinical setting.

In a study the following year by the Core Curriculum Task Force of the AAUAP's Training Director's Council, the utilization of state Title V clinics was studied. Of the 34 UAF Training Directors who responded, half reported that state funded clinics are used as training sites and the number of clinics employed varied from 11 clinics used by one UAF to the use of one clinic by six UAFs.

A question on interest in staff exchanges elicited 24 positive and 16 negative responses. Comments provided by respondents provided additional insight. One for example, noted that "a faculty in residence (during summer? or academic holidays?) in situ would be most productive. More local staff could participate this way." No mention was made of how this might be financed but another respondent commented that, "We would be interested in an exchange program where a staff member could go to a UAF or another program for on-job experiences. Support would be required as our state has frozen all out-of-state travel."

Funding was the subject of a question on the survey in which the state

programs were asked if their program had a budget for inservice training and, if so, what the state's plan provided relative to the frequency for training. The responses were about evenly split: twenty-one (21) said the program did budget for inservice training and twenty-two (22) responded negatively. Regarding the "schedule" or frequency for training, seventeen (17) reported that training was plannad for as the occasion arises and ten said that the program planned for training on a yearly basis. One respondent reported a plan for training every three years.

After the Survey

The survey results were reported in the "Survey of MCH and CC Training Needs - Summary of Results" which was distributed to all Directors of MCH and CC programs and to both the Directors and Training Coordinators of the UAFs in the summer of 1983. Recipients of the report within UAFs were encouraged in a cover letter to follow up with state Title V Directors to directly confirm or to update or expand upon the training needs for that particular state.

In addition to the obvious reasons for following up, there was a strong feeling on the part of some that the respondents had not uniformly reported training needs of local health department personnel, but had focused only on state level staff. Followup dialogue did occur in a number of instances and in some cases the results can be directly traced to relationships and activities which are continuing today and which have been reported in this publication and in its predecessor, Developmental Handicaps: Prevention and Treatment II.

Although the exact number is not known, a fair number of the proposals for continuing education projects funded through DMCH for FY '85 funding were the result of such liaisons. Many of those proposals cited the national survey of training needs in their background/needs statements. Ultimately, seven of eiwhteen projects awarded were for UAF-State Title V projects, of which two are recurring, multi-year projects.

In Ohio, the national survey was followed by a statewide assessment of MCH and CC training needs. In the latest step of that effort, a task force is working to develop a three-year plan for continuing education. Members of the task force include the Ohio Maternal and Child Health, the Nisonger Center UAF, and the University Affiliated Cincinnati Center for Developmental Disabilities UAF.

In a recent sampling of Training Directors of UAFs the Training Director's Council asked for an indication of the areas in which they could provide training. The responses, expressed in percentages, are shown in the following:

Areas in which UAF Training can be Provided

Interdisciplinary Team Functioning	100%
Family Support Services	94%
Genetics	75%
Screening for Handicaps	88%
Developmental Testing/Diagnosis	100%
Case Management	94%
Behavioral Management	94%
Early Intervention	94%
Fiscal Management	38%
Use of Statistics	56%
Evaluation of the Physically Handicapped Person	94%
Evaluation of the Mentally Retarded Person	100%
Effect of Chronic Illness on the Family (Emotional-Social-Economic)	88%
Influences of Cultural Differences on Management of Health Problems	44%

Summary and Some Thoughts for the Future

Certainly, training needs are continually evolving though the changes are not drastic. In terms of how to deal with and meet those needs, the implication is for strong continuing interaction and working relationships at the local level. National surveys such as the one discussed here cannot fulfill that role. The national survey can stimulate interaction at the level at which the work is carried out, and in the past it has been shown that there is a mutual desire on the part of the networks of UAFs and State MCH and CC programs to work together. That desire continues and provides the impetus to continue to seek mutual training opportunities that will enhance the delivery of services to the handicapped and disabled children and their families which the programs are committed to serve.

Surveys take time and resources which often might be better spent in providing the training and service rather than talking about it. Periodically, though, it is necessary to step back and collectively review the status of a subject. With the survey of MCH and CC program training needs now almost three years past, consideration must be given to looking ahead and planning how these networks can jointly project the course of their future alliances. Whether or not to survey again is not the central question, but rather, we must consider how to analyze trends and anticipate training needs of the future.

Acknowledgment:

The training needs of State MCH and CC Programs were studied by a subcommittee of the Association of Maternal and Child Health and Crippled Children's Programs in association with the MCH Consortium Project of the American Association of University Affiliated Programs (AAUAP). Special acknowledgment is given to Victor D. Menashe, M.D., who chaired the subcommittee. More than that, it was only through his efforts that the design of the survey and the survey process itself were completed. At the time, Dr. Menashe was Director of the Crippled Children's Division and the Child Development and Rehabilitation Center of the Oregon Health Sciences University. Thus, his role as Director of both a University Affiliated Facility and the state CC program, was pivotal. He well understood the unique relationship that UAFs and Title V programs can enjoy. His belief in the importance of those is reflected in the efforts he exerted with this study. More recently, he provided guidance in the preparation of this paper.

Notes and References

The "Survey of MCH and CC Training Needs: Summary of Results" is an unpublished report available by contacting the AAUAP, 8605 Cameron Street, Suite 406, Silver Spring, MD 20910.

The study referenced on page 103 is an unpublished report of the results of a survey conducted by Calvin Knobeloch, Ph.D., Training Director of the Division for Disorders of Development and Learning, The University of North Carolina-Chapel Hill, and Mary Ann Smith, Ph.D., Associate Director, Child Development Center, the University of Tennessee Center for Health Sciences, Memphis, Tennessee.

The Developmental Handicaps: Prevention and Treatment series is published by the AAUAP. The 1983 edition is now out of print. Copies of Developmental Handicaps: Prevention and Treatment II, published in 1984 may be obtained by contacting AAUAP, 8605 Cameron Street, Suite 406, Silver Spring, Maryland 20910.

A Summary of Papers Published in the Developmental Handicaps Series

1983:

Johnson, Nancy M. and Chamberlin, Harrie R., "Early Intervention: The State of the Art"

Kaminer, Ruth K. and Cohen, Herbert J., "Intellectually Limited Mothers"

Berg, Cynthia and Emanuel, Irvin, "Relationship of Prenatal Care to the Prevention of Mental Retardation and Other Problems of Pregnancy Outcome"

Ireys, Henry and Magrab, Phyllis, "Human Cost of Chronic Illness"

1984:

Burney, Linda R., Walker, Ann P. and Dumars, Kenneth W. "Prenatal Diagnosis: The State of the Art"

Ouellette, Eileen, "The Fetal Alcohol Syndrome"

Nelson, Richard P., "Adolescents with Chronic Illness or Disability"

Davidson, Phillip W. and Roghmann, Klaus J., "Methodology for Forecasting Service Needs for Chronically Ill and Developmentally Handicapped Persons"

Crocker, Allen C., "Regionalization of Health Service Systems"

1985:

Palmer, Frederick B., Ziring, Philip, and Shapiro, Bruce K., "Serious Viral Infection in Developmentally Disabled Persons: Threat to Normalization?"

Vanderheiden, Gregg C. and Dolan, Terrence R., "Promises and Concerns of Technological Intervention for Children with Disabilities"

McPherson, Merle G., "Community Based Services for Disabled/Chronically Ill Children and Their Families"

Meryash, David L., "The Fragile-X Syndrome"

Resource Information

For additional resources on these topics, the reader is referred to the "1985-86 Resource Guide to Organizations Concerned with Developmental Handicaps," which is published by the AAUAP. In the current edition, a cross reference listing of UAF resources is provided, indexed by topic area. To obtain a copy, contact the AAUAP.

Each year, a series of workshops is developed to accompany the topics of papers published in the series. Workshops which have been presented to date include:

"Early Intervention: The State of the Art," presented at the spring meeting of the Northwest Developmental Pediatric Society. Planning by the three Pacific Northwest UAFs--Child Development and Rehabilitation Center (Portland), Child Development and Mental Retardation Center (Seattle), the Center on Human Development (Eugene) and the Oregon Crippled Children's program and Idaho MCH and CC Programs.

"The Human Cost of Chronic Illness," presented in conjuction with a conference planned by the Georgetown University Child Development Center and the Virginia State Department of Health.

"Intellectually Limited Mothers," planned and co-sponsored by the Rose F. Kennedy Center UAF and the New York State Department of MCH and CCS and the New York City Department of MCH and CCS (The Bureau of Handicapped Children).

"Intellectually Limited Mothers," planned in association with the conference on high risk parenting with planning by the Developmental Evaluation Center and Eunice Kennedy Shriver Center UAFs, Massachusetts Departments of Public Health, Social Services and Mental Health, and the Region I Maternal and Child Health Office.

"The Relationship of Prenatal Care to the Prevention of Mental Retardation and Other Problems of Pregnancy Outcome," at the Second Annual March of Dimes Medical Symposium. Planning groups included the Mental Retardation Institute UAF, the NY Maternal and Child Health Program, Westchester County Department of Health, March of Dimes Chapters (Westchester, Rockland and Putnam Counties), Lienhard School of Nursing of Pace University, and the Westchester County Medical Center's Department of Obstetrics and Gynecology.

"The Relationship of Prenatal Care to the Prevention of Mental Retardation and Other Problems of Pregnancy Outcome," planned and conducted by the Department of Pediatrics, University of Kentucky Medical Center, The Kentucky Division of Maternal and Child Health, and the Human Development Program UAF (Lexington).

"Fetal Alcohol Syndrome," presented by the Georgetown Child Development Center and the District of Columbia Crippled Children's Services and Administration for Drug and Substance Abuse.

"Coping with Chronic Illness in Adolescence: Personal Experiences and Professional Perspectives," planned and presented by the Center for Developmental Disabilities UAF at the University of Vermont, the Vermont Department of Health, United Cerebral Palsy of Vermont, the Epilepsy Association of Vermont, and the Vermont Center for Independent Living.

"Alcohol and Pregnancy," two of four regional workshops planned and presented by the Pennsylvania Department of Health, Division of Maternal and Child Health and the Temple University Developmental Disabilities Program.

"Awareness of Service Resources for Children with Special Needs and Increasing Awareness of the South Dakota Fetal Alcohol Task Force," presentations of the South Dakota University Affiliated Center for Developmental Disabilities and the South Dakota Maternal and Child Health and Crippled Children's Handicapped Services Programs as part of the South Dakota Public

Health Association meeting.

A teleconference on Ohio's "Program for Prevention of Developmental Disabilities" to illustrate the Ohio coordinated network of prevention education and public awareness activities planned and presented by a consortium including: Health Education Alliance through the University of Cincinnati, the Division of Maternal and Child Health of Ohio, the Nisonger Center and University Affiliated Cincinnati Center for Developmental Disabilities UAFs, the Ohio State University Area Health Education Centers, and the Ohio Cooperative Extension Service.

"Fetal Alcohol Syndrome," planned by the Montana University Affiliated Program Satellite and the Montana Maternal and Child Health and Crippled Children's Services.

"Human Cost of Chronic Illness," followup to a 1984 conference developed by the Georgetown University Child Development Center and the Division of Family Health of Virginia.